Presentation, Radiologic Features, and Treatment Options of Congenital Tongue Tumors: A Comprehensive Review

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AIM: Congenital tumors of the tongue are rare in pediatric patients but encompass a diverse range of entities. Each tumor type exhibits distinct clinical behaviors, necessitating a precise approach to differentiating the tumor types and a tailored, tumor-specific treatment regimen. Advanced imaging techniques, such as diffusion-weighted imaging and perfusion studies, play a vital role in differentiating benign and malignant tongue tumors. This review summarizes current knowledge regarding the presentation, imaging features, and treatment of congenital tongue tumors.

METHODS: A literature review was conducted by searching studies on congenital tongue tumors in databases such as PubMed, Embase, Web of Science, and Scopus. Relevant data, such as clinical features, radiologic characteristics, treatment modalities, and outcomes for different tumor types, were extracted from the selected articles.

RESULTS: Our literature review reveals the various entities of congenital tongue tumors, which can be categorized in terms of hereditary pattern, phenotype, and rarity. Congenital tongue tumors include a range of vascular malformations, such as hemangiomas, lymphatic malformations, arteriovenous malformations, and venous malformations. Another entity is represented by cystic lesions, including dermoid cysts, epidermoid cysts, ranulas, and mucous retention cysts. Rare malignant neoplasms include teratomas and rhabdomyosarcomas. These tumor types vary in terms of swelling, respiratory distress, or impaired oral function, depending on size and location. The detection of these tumors can be carried out using imaging modalities, such as ultrasound, magnetic resonance imaging, and computed tomography, which are utilized to facilitate diagnosis and differentiation. At present, surgical excision remains the cornerstone of treatment, while other modalities may be adopted, depending on tumor type and extent. The prognosis of congenital tongue tumors can be affected by tumor's site, size, involvement of vital structures, and malignancy.

CONCLUSIONS: Given their diversity and complexity, congenital tongue tumors, albeit uncommon, require specialized clinical treatments tailored to each tumor type's characteristics. Understanding the variable presentations and imaging features enables accurate diagnosis, while customized treatment strategies are key to optimizing outcomes and minimizing morbidity in pediatric tongue tumors. This review summarizes current knowledge aimed at enhancing differential diagnosis and management of these diverse entities.

Keywords: congenital tongue tumors; pediatric patients; tailored approach; diagnosis and management; imaging features

Introduction

Congenital tongue tumors, albeit rare, represent a captivating facet of pediatric pathology. The overall incidence of congenital tongue tumors varies across different studies and populations, with reported rates ranging from 1.7 to 13.5 per 100,000 births [1]. These lesions occur during childhood and affect both males and females equally. It is known that the classifications of congenital tongue tumors are highly diverse. For instance, Yuhan *et al.* [2] have documented up to 65 soft tissue tumors that affect the tongue, with hemangiomas and lymphovascular malformations being the most prevalent types, accounting for 80% of the cases. Several entities, such as teratomas, hemangiomas, lymphatic malformations, and tongue fibromatosis, present unique challenges in diagnosis and management [3]. Besides, it should be highlighted that prenatal detection of malignant tongue tumors has been reported [4]. The clinical presentation of congenital tongue tumors spans a spectrum of signs and symptoms, including visible swelling, dysphagia, and, in

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severe cases, respiratory distress. In addition, congenital tongue tumors may lead to various complications, including feeding difficulties and speech impairments, highlighting the importance of early diagnosis and intervention [2, 3, 4]. Beyond their rarity, these tumors exhibit distinctive clinical behaviors, necessitating a precise approach to differential diagnosis and therapeutic interventions tailored to different tumor types [5].

Imaging modalities, such as ultrasound, magnetic resonance imaging (MRI), and computed tomography (CT), play pivotal roles in unraveling the radiologic features of congenital tongue tumors, providing insights into their extent, composition, and potential impact on adjacent structures, which represent a set of essential information to aid diagnosis and therapy planning [6]. However, it should be considered that general anesthesia may be necessary in some situations. If the child is too young to cooperate or has difficulty remaining still, or if the MRI examination requires a longer duration that the child cannot tolerate while awake, sedation anesthesia is required to ensure that the child remains completely still and comfortable throughout the procedure, allowing for high-quality imaging [6, 7]. Because of its real-time nature and the non-ionizing radiation involved, ultrasound is a potent technique for initial screening [7], providing crucial information regarding the size, location, and vascularity of a tumor. However, more sophisticated modalities, such as MRI and CT, may be needed in cases hampered by limited soft tissue contrast and the inability to evaluate deep structures [2]. While CT is excellent at identifying bone involvement, MRI offers a more complete assessment of the tumor's size and connection to surrounding tissues due to the higher resolution afforded to image soft tissue [8, 9, 10]. Owing to the diverse histological subtypes, the congenital tongue tumors do not necessarily all exhibit the same radiologic appearance. Typically, hemangiomas appear on contrast-enhanced images as well-defined, aggressively enhancing lesions, but lymphatic malformations may present a more complex appearance with fluid-fluid levels and septations [11]. The development of sophisticated imaging techniques, such as diffusion-weighted imaging and perfusion studies, has gradually expanded the items in the diagnostic toolbox for congenital tongue tumors, providing aids to distinguish benign entities from malignant ones and offering important insights into the biological behaviors of these tumors [12]. Similarly, the therapeutic landscape for congenital tongue tumors is inherently intricate, but surgical excision remains the cornerstone in the therapeutic management of the disease [6], particularly for localized, well-circumscribed lesions [13]. However, the choice of treatment modality is contingent upon the type and size of tumor, as well as the potential implications of treatment for vital structures. Other treatment options may be taken into consideration in cases when surgical excision is not feasible or could cause considerable morbidity [14, 15, 16]. For example, sclerosing therapy, which involves injection of a sclerosing agent into the tongue tumor, has demonstrated promise in the treatment of some vascular malformations, including venous and lymphatic abnormalities [17]. Another popular therapeutic approach for treating vascular tongue tumors, especially arteriovenous malformations (AVMs), is embolization [17, 18]. Through the selective blockage of the feeding veins that support cancer growth, this minimally invasive treatment causes the tumor to reduce vascularization and eventually recede. Various materials, such as coils, particles, or liquid agents, can be used for embolizing tongue lesions, depending on the desired degree of blockage and the unique characteristics of the lesion. The distinct features of every tumor and the intended therapeutic outcomes—which range from symptom relief to total tumor eradication—determine which of these modalities should be used [19]. The long-term effects on the patient's quality of life must be considered while weighing the possible hazards and advantages of each strategy [20]. Furthermore, a multidisciplinary effort involving pediatric surgeons, otolaryngologists, radiologists, and oncologists is imperative to formulating comprehensive and patient-centric management plans. This review aims to provide a scholarly exploration of the clinical presentation, radiologic characteristics, and contemporary treatment paradigms of congenital tongue tumors. By consolidating current knowledge and fostering an understanding of these rare conditions, we aspire to contribute to the broader landscape of pediatric oncology and promote advancements in the diagnosis and treatment of congenital tongue tumors.

Methods

Literature Search Strategy

A comprehensive literature search was conducted to identify relevant studies on congenital tongue tumors in pediatric patients. No year restrictions were applied during the article retrieval process to ensure a comprehensive analysis. Articles were retrieved from four major electronic databases, namely PubMed, Embase, Web of Science, and Scopus. The search strategy included the use of a combination of Medical Subject Headings (MeSH) terms and keywords related to congenital tongue tumors, such as "congenital tongue tumor", "pediatric tongue tumor", "lingual tumor", "hemangioma", "lymphangioma", "teratoma", and "rhabdomyosarcoma". Studies were considered eligible for inclusion if they met the following criteria: (1) presented in the form of original research articles, case reports, case series, or reviews focusing on congenital tongue tumors in pediatric patients (aged 0–18 years); (2) we did not adopt any language restriction; and (3) provided information on the clinical presentation, radiologic features, treatment modalities, or outcomes of congenital tongue tumors. Studies were excluded if they (1) focused on adult patients or acquired tongue tumors; and (2) did not provide sufficient information on the outcomes of interest.

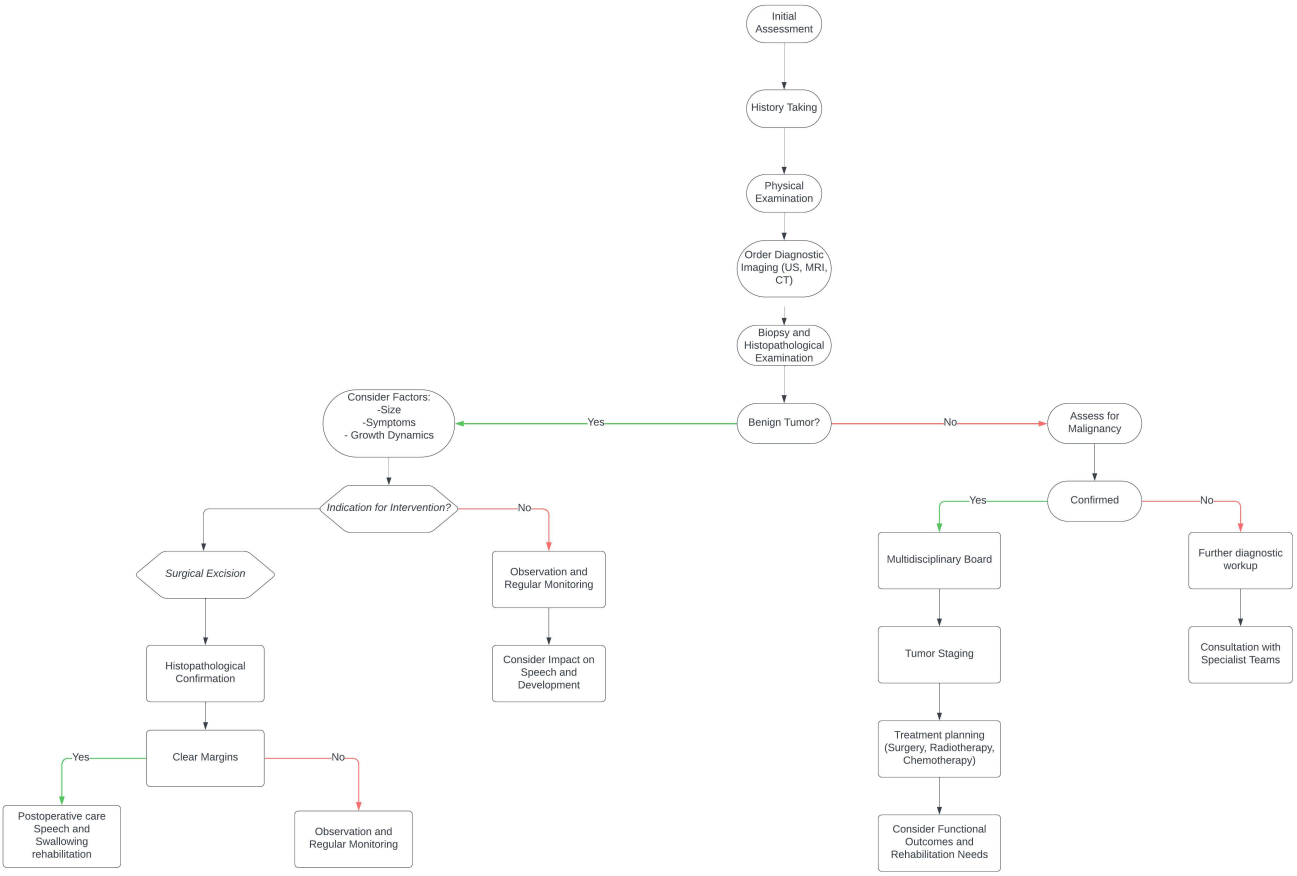

Fig. 1. Flowchart depicting the management of tongue tumors in pediatric patients (Paint-3D, Microsoft Corporation, v.6.19, 2024, Redmond, Washington, D.C., USA). US, ultrasound; MRI, magnetic resonance imaging; CT, computed tomography.

Study Selection and Data Extraction

The initial database search yielded a total of 331 records. After removing duplicates, the titles and abstracts of the remaining studies were screened independently by two reviewers (AM and LLV) to assess their eligibility based on the inclusion and exclusion criteria. Any discrepancies were resolved through discussion with a third reviewer (SL) until a consensus was reached. The full texts of the potentially eligible studies were then retrieved and reviewed in detail to determine their final inclusion in the review. Due to the heterogeneity of the included studies and the lack of standardized reporting, a meta-analysis was not feasible. Instead, a narrative synthesis of the findings was conducted, focusing on the clinical presentation, radiologic features, treatment modalities, and outcomes of congenital tongue tumors in pediatric patients. The results were organized by tumor type and presented in tables and figures where appropriate. Research involving human subjects, human materials or human data were conducted in accordance with the Declaration of Helsinki. Informent consent was obtained for each patients' data included.

Results

Congenital tongue tumors comprise a diverse spectrum of entities, encompassing dermoid cysts, lymphangiomas, teratomas, and hemangiomas. The tumors exhibit unique characteristics such as pathogenesis and clinical features, and are epidemiologically different. Furthermore, the diagnostic approach, management, and prognosis vary from one tumor type to another. This comprehensive overview aims to delve into the distinctive aspects of each congenital tongue tumor, offering insights into their nature and providing a foundation for understanding their varied presentations, diagnostic approaches, and management strategies (Fig. 1).

Table 1 summarizes the key attributes of congenital tongue tumors, offering a comparison of their specific features.

Vascular Malformation

Vascular malformations refer to abnormal formations of blood vessels within the respiratory system. These malformations affect not only the tongue but also various components of the airway, including the trachea, bronchi, and lungs [6, 7, 8]. Vascular malformations are typically characterized by enlarged, tangled, or malformed blood vessels, which may lead to functional and structural abnormalities

Table 1. Summary of main congenital tongue lesions, clinical features, and management.

Tumor type	Age of presentation	Clinical presentation	Associated syndrome	Radiological features	Treatment	Outcome
Hemangiomas	Childhood	Visible swelling, dysphagia, respiratory distress	None	US: Well-defined, homogeneous, hypervascularity	Propranolol, corticosteroids, surgical excision	Generally favorable; watch-and-wait for regression
Arteriovenous malformations	Variable	Asymptomatic to locally destructive and infiltrative entities	Hereditary hemorrhagic telangiectasia	Angiography: Abnormal connections, hypertrophy, nidus	Embolization, surgical excision, targeted medical therapy	Recurrence; comprehensive treatment needed
Venous malformations	Present at birth	Dysphagia, sleep apnea, bleeding, macroglossia, speech dysfunction, pain, disfigurement	None	US: Solid, hyperechoic, transcends normal planes	Conservative approaches, surgical resection, laser therapy, sclerotherapy	Variable; cautious approach due to risks
Lymphatic malformations	Childhood	Hemorrhage, excessive salivation, speech disturbances, airway obstruction	None	US: Predominantly cystic masses, fluid-fluid levels	Excision, cryotherapy, electrocautery, sclerotherapy, corticosteroid administration	Functional and aesthetic challenges in some cases
Dermoid and epidermoid cysts	Present at birth	Painless, slow-expanding masses	None	US or MRI: Cystic nature, anatomical relationships	Surgical excision	Typically benign; recurrence prevention
Mucous retention cysts	Present at birth	Painless, fluid-filled swellings	None	Clinical examination, US or MRI	Conservative surgery for symptomatic cases	Generally asymptomatic; surgical intervention if needed
Teratomas	Infancy	Tongue displacement, potential protrusion from the oral cavity	Trisomy 13, Aicardi syndrome, Pierre Robin syndrome	US: Cystic-solid mass, calcification, blood vessels from palate or tongue	Early and complete excision	Benign, but potential for malignancy with age
RMS	Childhood	Tongue as atypical site; may include painless, slow-expanding masses	None	T1-weighted MRI: Hypo- to isointensity; T2-weighted MRI: Hyperintensity	Surgery, chemotherapy, radiation therapy	Depends on factors such as tumor size and location

Abbreviations: US, ultrasound; MRI, magnetic resonance imaging; RMS, Rhabdomyosarcoma.

in the airway. There are different types of vascular malformations, with distinctive characteristics determined by factors such as size, location, and extent of the malformation [14, 15, 16].

Hemangiomas

Hemangiomas represent the most prevalent tongue tumors during childhood, occurring in approximately 4–5% of infants [10, 11]. Hemangiomas can be classified as infantile or congenital. The infantile subtype is characterized by a rapid proliferation phase until the infant reaches 12 months of age, followed by a gradual involuting phase that occur between 6 and 9 years of age [7]. Congenital hemangiomas, while less common, are more likely to be detected in prenatal imaging. This subtype undergoes proliferative phase *in utero*, manifesting full size at birth, and can subsequently remain stable, partially resolved, or fully involute within the first year of life [8]. These two subtypes can be pathologically differentiated from each other based on the glucose transporter 1 (GLUT-1) expression and endothelial cell turnover. Infantile tongue hemangiomas express GLUT-1 and exhibit increased endothelial cell turnover, while congenital hemangiomas are GLUT-1-negative lesions composed of capillary lobules and endothelial cells, often associated with large veins, arteries, and lymphatic vessels [9]. Congenital tongue hemangiomas that do not undergo a regression phase and growing in proportion with the child's development are referred to as non-involuting congenital hemangiomas, which exhibit prominent coarse telangiectasia and peripheral blanching [10, 11]. Mucosal hemangiomas are raised and red, while submucosal variants appear blue or purple. Hemangiomas, particularly those extensively involving the lip and tongue, can lead to macroglossia, which, when circumscribed within the tongue, can be differentiated from a more systemic process based on the circumscribed vascular nature of the mass [12]. Despite their distinct characteristics, a watch-and-wait strategy is commonly applied to both subtypes to achieve spontaneous regression. Active intervention is deemed necessary when complications arise, such as uncontrolled hemorrhage, pain, ulceration, infection, or airway obstruction. For oral cavity hemangiomas, medications such as propranolol are administered as the first-line treatment. Beta-blockers, particularly propranolol, have emerged as a first-line treatment option for infantile hemangiomas, with studies demonstrating its therapeutic potential in size reduction and inducing a color shift from bright red or purple to a duller red or gray of the mass during the proliferative phase [13]. Alongside propranolol, oral or systemic corticosteroids have proved effective in many cases. Surgical excision, carbon dioxide laser treatment, cryosurgery, and sclerosing agents are additional modalities employed in managing larger oral cavity hemangiomas [14]. However, surgical excision of hemangiomas may be associated with complications such as bleeding, infection, and scarring, underscoring the need for more prudent patient-surgery matching and more meticulous pre-operative planning [13, 14, 15, 16]. Hemangiomas can also be detected in fetus during prenatal ultrasound examinations; therefore, postnatal MRI examination is required for comprehensive anatomic characterization of the mass after child delivery. Anatomically, hemangiomas and other vascular anomalies exhibit a lack of adherence to the typical boundaries of oral and neck spaces—a conspicuous manifestation of trans-spatial location (Fig. 2a,b). The ultrasound approach is encumbered with limitations in evaluating the complete anatomic extent of hemangiomas. According to the ultrasound findings, hemangiomas of the tongue typically present as well-defined, homogeneous, solid masses with varying echogenicity, and demonstrate hypervascularity with potential arterial and venous waveforms [15]. Thus, the MRI lends itself useful in accurately defining the extent of hemangiomas. Deep trans-spatial hemangiomas appear as solid masses with intermediate signal intensity on T1-weighted MRI and display heterogeneous hyperintensity on T2-weighted MRI. In postnatal MRI, these masses typically display homogeneous avid enhancement. Additionally, associated features such as dilated vasculature or flow voids in signal intensity may be observed. Importantly, tongue hemangiomas may remodel adjacent bone but do not invade osseous structures [16].

Arteriovenous Malformations

AVMs can occur in various head and neck regions, including the face, mouth, jaw, and neck tissues. Although considered congenital, AVMs may not become symptomatic until later in life [17]. This kind of malformation is characterized by high-flow vascular anomalies in which there exist abnormal connections between arteries and veins that result in a continuous shunting of blood across a defective capillary bed. This pathogenic process leads to the dilation and arterialization of postcapillary venules and hypertrophy of feeding arteries, forming a "nidus" [18].

AVMs have an extensive spectrum of presentations, ranging from asymptomatic minor lesions to highly aggressive, locally destructive, and infiltrative entities [19]. Recognized as the most perilous subtype of vascular malformations, AVMs pose a significant threat due to their aggressive behavior that potentially puts patients in a life-threatening state. The extent of AVMs is a crucial factor in their characterization, distinguishing between focal and diffuse forms [20]. Focal AVMs typically feature more defined margins, involve one or two anatomic subsites, and are minimally infiltrative, while diffuse AVMs exhibit ill-defined margins, multiple feeding vessels, and invasive characteristics. The International Society for the Study of Vascular Anomalies (ISSVA) categorizes AVMs into four classifications, namely sporadic, associated with hereditary hemorrhagic telangiectasia, associated with capillary malformation, and others [21]. AVMs exhibit distinct imaging characteristics across various modalities (Fig. 3a,b). The Schobinger stag-

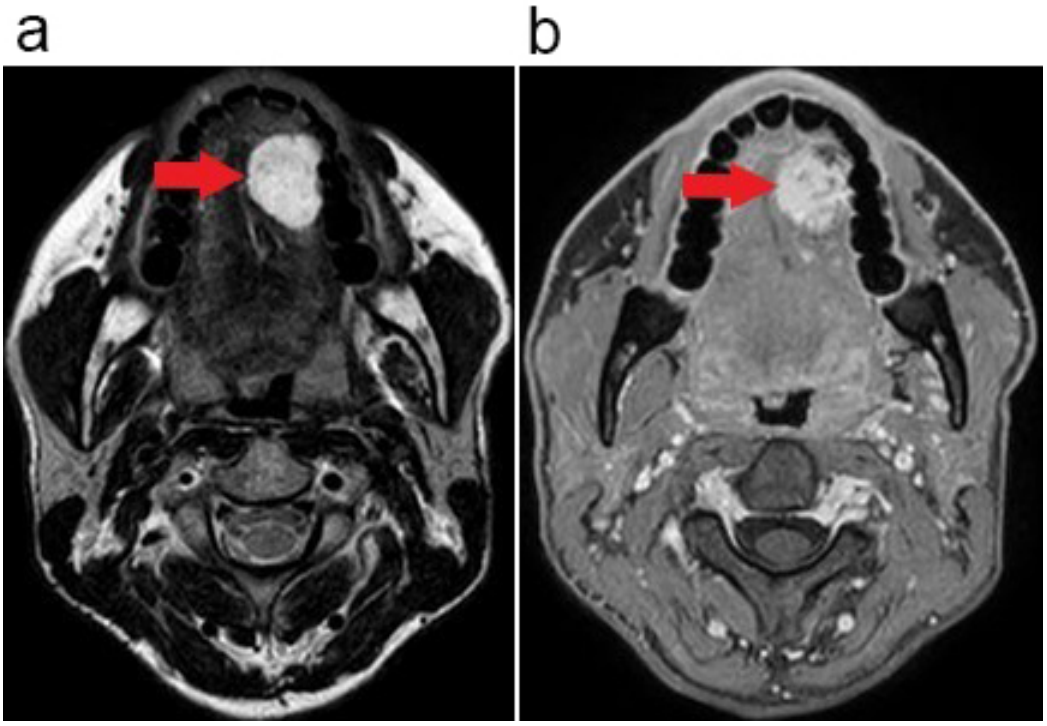

Fig. 2. MRI findings of pediatric hemangioma of oral cavity. Axial T2- weighted (a) and T1-weighted fat-saturated post-contrast (b) MRI images showing an oval-shaped formation located at the body-apex of the tongue characterized by marked hyperintensity on T2 and showing progressive contrast enhancement, attributable to an hemangioma (Paint-3D, Microsoft Corporation, v.6.19, 2024, Redmond, Washington, D.C., USA). Red arrows indicate the border of lesion. This image is from a patient at University hospital of Catania and the study was performed with the patient's consent.

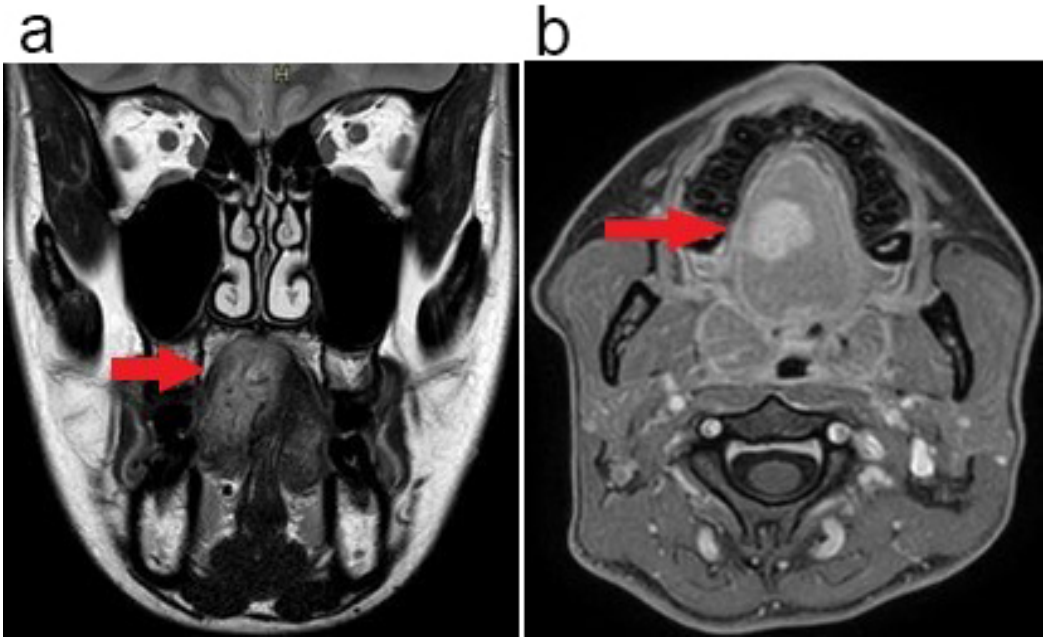

Fig. 3. Radiologic examination of an arteriovenous malformation. Coronal T2-weighted (a) and axial T1-weighted (b) fat saturated post-contrast MRI images showing a oval-shaped lesion with clear margins, which distorts the upper profile of the tongue, and centrally depicting altered signal intensity, hyperintense on T2 sequences with some serpiginous signal voids, in addition to homogeneous, intense, and persistent contrast enhancement. Within the voids, ectatic vascular structures are recognizable (Paint-3D, Microsoft Corporation, v.6.19, 2024, Redmond, Washington, D.C., USA). Red arrows indicate the border of the lesion. This image is from a patient at University hospital of Catania and the study was performed with the patient's consent.

ing system utilizing a clinical progression-based approach is commonly employed for AVMs [22]. AVMs are relatively rare vascular malformations, with intracranial AVMs being significantly more prevalent than their extracranial counterparts, accounting for nine in every ten AVM cases [23]. Extracranial AVMs have been identified in a total of 3.7–4.7% of patients [24, 25], although the exact incidence remains unknown, mostly affecting the head and neck regions [26].

Treating AVMs of the tongue presents a complex challenge, given their invasive and destructive nature. Adding to the treatment challenge is the recurrence, which is common in AVMs. Therefore, a comprehensive, multidisciplinary treatment approach is essential for improving patient outcomes [27, 28]. Both embolization and surgical excision are the mainstay of AVM treatment, with targeted gene-based therapy emerging in the treatment landscape. Although sclerotherapy can be considered as an alternative treatment strategy for malformations, this form of treatment carries risks such as postprocedural thromboembolism and life-threatening tongue swelling, which necessitate cautious and close post-treatment monitoring [29, 30, 31]. Successful surgical interventions, with or without embolization, require the removal of the nidus [32]. In cases where larger, focal, or diffuse lesions involving multiple anatomic areas, surgical reconstruction is often required [33]. Considered the gold-standard detection strategy, angiography enables detailed visualization of the abnormal connections, or "nidus", between arteries and veins, showcasing hypertrophy of feeding arteries and arterialization of postcapillary venules. MRI reveals intermediate signal intensity on T1-weighted images, heterogeneous hyperintensity on T2-weighted images, and homogeneous avid enhancement post-contrast. Moreover, magnetic resonance angiography (MRA) offers non-invasive, three-dimensional imaging of blood vessels, whereas computed tomography angiography (CTA) provides three-dimensional visualization, capturing dynamic contrast enhancement in both arterial and venous phases, aiding in the identification of the nidus and abnormal vessel connections. Ultrasound is used to evaluate blood flow and distinguish between solid and cystic components [34]. Periodic radiologic examinations, including CTA and ultrasonography, are vital for the close follow-up and early detection of recurrence, improving management and treatment planning for AVMs [35].

Venous Malformations

Venous malformations (VMs) of the tongue are a rare but clinically significant vascular anomaly in children, accounting for approximately 40% of all diagnosed vascular anomalies. They affect males and females equally [36]. These malformations involve abnormal development of veins within the tongue, leading to localized vascular abnormalities [37]. Addressing tongue VMs in children presents a multifaceted challenge due to their diverse clinical manifestations, encompassing dysphagia, sleep apnea, bleeding, macroglossia, speech dysfunction, pain, and disfigurement [12]. The management of these lesions involves navigating various therapeutic options. In cases of asymptomatic VMs, conservative approaches or observation may suffice, while symptomatic cases necessitate interventions such as surgical resection, laser therapy, sclerotherapy, and hybrid therapies. Caution is exercised in patients with multifocal or large-volume VMs, as they carry an elevated risk of localized intravascular coagulopathy. Assessing D-dimer levels, prothrombin times, partial thromboplastin times, and fibrinogen levels is integral to lesion management due to the increased risk for spontaneous intralesional thrombosis [38]. Although the risk of postprocedural thromboembolism in head and neck VM treatments is rare, the utility of perioperative anticoagulation is controversial [39]. Interventional risks concerning the oral cavity include perioperative edema and airway obstruction, with documented instances of life-threatening tongue swelling after sclerotherapy. Hence, a cautious approach with a low threshold for inpatient observation is needed. While surgical resection ensures a definitive cure, the associated morbidity, particularly when extensive tongue tissue is involved, underlines the line to explore less invasive alternatives, such as neodymium-doped yttrium aluminum garnet (Nd:YAG) laser system. Nd:YAG laser system offers a nuanced balance between elevated efficacy and reduced morbidity in the treatment of oral mucosal VMs [40]. Contrary to hemangiomas, VMs are typically diagnosed based on clinical evaluation rather than solely on imaging findings [41, 42, 43]. Cross-sectional imaging is particularly valuable for assessing the anatomic extent of VMs. The ultrasound reveals a solid hyperechoic mass that is often compressible and transcends the normal planes within the oral and neck spaces. Identifying shadowing phleboliths during ultrasound boosts diagnostic confidence, especially in recognizing the venous subtype of malformation. Doppler flow in VMs may present as slow, monophasic, or absent [15]. In MRI, VMs may exhibit similarities to hemangiomas, presenting with T1-weighted intermediate signal intensity and heterogeneous T2-weighted hyperintensity; flow voids are less frequently observed. Enhancement, though common, is more evident in delayed gadolinium-enhanced MRI of VMs compared to postnatal hemangiomas [44]. Distinguishing features for VMs include calcified phleboliths, which are best visualized using gradient-echo sequences or as T2-weighted hyperintense "venous lakes", though their presence is not universally noted [16].

Lymphatic Malformations

Lymphatic malformations (LMs), characterized by vascular anomalies of unknown origin, constitute approximately 6% of all benign soft-tissue tumors in children [45]. In pediatric Head and Neck (H&N) patients, lymphatic malformations may exhibit long-term effects on craniofacial development,

emphasizing the importance of early intervention and multidisciplinary management [46, 47, 48]. The occurrence of LMs in the tongue can give rise to a spectrum of symptoms, including hemorrhage, excessive salivation, speech disturbances, chewing and swallowing difficulties, airway obstruction, and orthodontic abnormalities like mandibular prognathism and malocclusion. The impact on the quality of life for individuals with LMs in the tongue is considerable, causing functional impairment and cosmetic deformities [46]. Various treatment modalities, including complete excision, cryotherapy, electrocautery, sclerotherapy, corticosteroid administration, and embolization, have been explored [47, 48, 49]. However, despite the range of available treatments, LMs of the tongue continue to pose functional and aesthetic challenges for patients. LMs can be distinguished from VMs and hemangiomas by means of ultrasound, with the former predominantly presenting themselves as cystic masses rather than solid structures and confining any observed flows to thin septa. Microcystic lesions may lack individual, discernible cysts and may appear echogenic. The presence of fluid levels and debris in the ultrasound may suggest lesional hemorrhage [15]. Importantly, the ultrasound characteristics of LMs can effectively predict their appearance on MRI. MRI plays a crucial role in characterizing LMs, distinguishing them from other vascular anomalies, and guiding treatment planning. A cystic LM exhibits a simple fluid signal intensity on MRI unless complicated by hemorrhage, where fluid-fluid levels become evident. Microcystic lesions may manifest as infiltrative T2-weighted hyperintensity. Contrast-enhanced postnatal imaging reveals minimal septal enhancement in cases where characterization is feasible. This distinct imaging profile aids in the accurate differentiation of LMs from other vascular anomalies [50].

Cystic Lesions

Congenital cystic lesions in the oral cavity are uncommon developmental anomalies that present as fluid-filled sacs or cavities within the oral tissues and are present at birth [51, 52, 53, 54, 55]. These lesions vary in size, location, and histological characteristics, encompassing a range of entities such as ranulas, dermoid cysts, epidermoid cysts, and mucous retention cysts [56, 57, 58, 59, 60]. Among these, cystic lesions of the pediatric tongue, while less frequent than vascular anomalies, still constitute a significant proportion of congenital tongue lesions [61]. Cystic lesions of the pediatric tongue typically show up on CT scans as well-defined, hypoattenuating formations with distinct borders in relation to the surrounding muscle tissue. They can be distinguished from more vascular lesions by their lack of contrast enhancement. However, CT is rarely utilized for assessments in young children due to concerns about radiation exposure. On the other hand, MRI offers a better contrast resolution, which is very helpful when assessing soft tissue features like the tongue. Due to the fluid content of the cysts, cyst lesions on MRI frequently exhibit a high signal intensity on T2-weighted imaging and a low signal intensity on T1-weighted imaging. Furthermore, MRI is a better option for evaluating pediatric patients because ionizing radiation is not involved. In the event that intervention is required, this modality also enables a thorough evaluation of the lesion's connection to the surrounding structures, which is essential for surgical planning (Fig. 4a,b).

Mucoceles, which are mucous retention cysts, are the most common cystic lesions of the oral cavity in children, with a reported incidence rate ranging from 0.2 to 0.9 per 1000 individuals [62, 63, 64]. Among all head and neck cysts, dermoid cysts—another type of congenital cystic lesion—have an estimated incidence of 1.6% to 6.9%, with the oral cavity being the second most common location of occurrence after the periorbital region [62]. The presence of these cystic lesions in the tongue can lead to various clinical manifestations, depending on their size and location, and may require surgical intervention for definitive management.

Ranula

A congenital ranula is characterized as a cystic malformation within the oral cavity, arising from fluid accumulation. This condition can result from the disruption of minor salivary ducts, leading to the extravasation of mucus into nearby structures that lack epithelial lining. Alternatively, it may stem from a blocked duct caused by atresia, osteal adhesion, or trauma. In cases of ductal obstruction, proximal expansion occurs, forming a mucus retention cyst that is typically observed in neonates [51]. The salivary gland system is heavily implicated in the development of the ranula cyst due to the presence of salivary duct epithelium lining it, which is most likely the result of duct obstruction or malformation. Moreover, the notion of a multiple etiology for congenital ranulas emphasizes how intricate this disorder is. It implies that a range of variables, including genetic and environmental ones, may interfere with the complex processes involved in the development of the oral cavity, resulting in these distinct deformities [52].

While congenital ranulas are often asymptomatic, causing minimal discomfort or functional disturbance, larger lesions can lead to difficulty in swallowing or speaking. Ranulas are classified into two main types: simple and plunging. A simple ranula is confined to the sublingual space, whereas a plunging ranula extends beyond the mylohyoid muscle into the neck [53]. Simple ranulas are typically asymptomatic, but it is important to note that they can potentially lead to airway obstruction [54]. Clinically, a ranula presents as a painless, bluish cyst beneath the tongue, often causing swelling that elevates the tongue and remains unchanged in size during activities like chewing, eating, or swallowing [52]. The optimal treatment approach for ranulas is still a subject of debate within the medical community. Some advocate for early marsupialization to mitigate potential complications such as sialadenitis, while others pro-

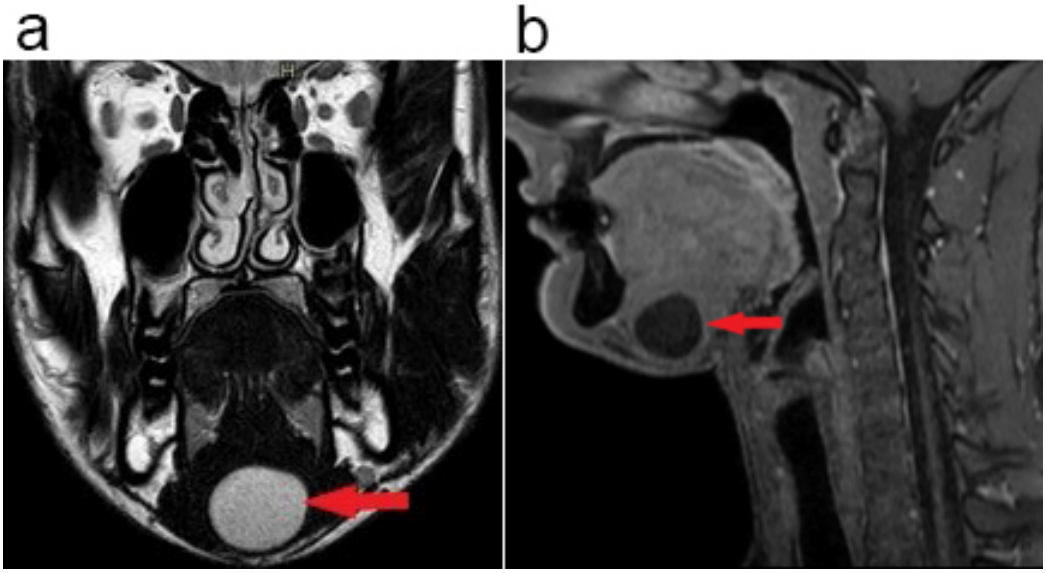

Fig. 4. Radiologic examination of an epidermoid cyst. Coronal T2-weighted (a) and sagittal T1-weighted (b) fat-saturated post-contrast MRI images showing an expansive formation with clear margins located in the middle of the floor of the mouth, between the digastric, mylohyoid, and geniohyoid muscles, which are imprinted and displaced laterally. The cyst contains dense liquid content with modest wall enhancement, without intracystic vegetative components (Paint-3D, Microsoft Corporation, v.6.19, 2024, Redmond, Washington, D.C., USA). The red arrows indicate the limit of the lesion. This image is from a patient at University hospital of Catania and the study was performed with the patient's consent.

pose a wait-and-see strategy, reserving surgical intervention for cases where issues such as airway obstruction or feeding difficulties emerge. In neonates, imperforate ducts may spontaneously resolve with feeding, and surgical interventions like needle aspiration, ranula excision with or without ipsilateral sublingual gland removal, marsupialization, cryosurgery, and sclerotherapy are among the available options. Given the tendency of congenital cysts to either resolve or rupture on their own, uncomplicated cases should be closely observed for potential spontaneous resolution over several months. The goal of treatment is to alleviate discomfort, prevent complications such as infection, and address any impact on oral functions [55].

The crucial radiological aspect in diagnosing a ranula, particularly when the cysts are sizable and have extended some distance from their origin, is identifying a connection to the sublingual space. This connection may manifest as a slender tail of fluid or a notable local fluid collection [56]. A simple ranula is situated within the sublingual space above the mylohyoid muscle, whereas a plunging ranula extends into the submandibular space, often accompanied by a collapsed sublingual portion referred to as the "tail". These distinctive features are essential for accurate diagnosis and aid in differentiating between simple and plunging ranulas. Radiologic means are applied to differentiating between simple and plunging ranulas. Since ranulas indifferently appear as cystic masses on ultrasound [57] and low-density lesions on CT scans [58], irrespective of their classifications, high signal intensity alteration on T2-weighted MRI images should be considered in differential diagnosis, to assist with distinguishing simple from plunging ranulas [59].

Dermoid and Epidermoid Cysts

Congenital dermoid and epidermoid cysts of the tongue are developmental anomalies characterized by cystic formations arising from embryonic remnants. Based on epidemiological data, dermoid cysts are the most common congenital cystic lesions of the tongue, followed by epidermoid cysts and ranulas. These cysts have the potential to develop in various anatomical locations throughout the body, and their occurrence rate in the head and neck area ranges from 1.6% to 6.9%, while their occurrence within the oral cavity specifically is estimated to be around 1.6% [60]. Despite their relatively common occurrence in the head and neck region, these cysts constitute a rare subset of oral cavity cysts, accounting for less than 0.01% of the total cases.

Dermoid and epidermoid cysts are often present at birth and arise from ectodermal elements during embryogenesis. Dermoid cysts typically contain skin, hair, and occasionally sebaceous and sweat glands, while epidermoid cysts are filled with keratinaceous material. The cysts are benign slow-growing masses, commonly found in the midline structures of the head and neck, including the oral cavity [61]. Clinically, they manifest as painless, slow-expanding masses that may be palpable or visible beneath the skin. Diagnosis is usually confirmed through imaging studies, such as ultrasound or MRI, which can reveal the cystic nature

and anatomical relationships [62]. Treatment for dermoid and epidermoid cysts involves surgical excision to prevent recurrence and address any cosmetic or functional concerns associated with the cyst [63]. Although rare, certain cystic lesions, such as dermoid cysts, have the potential for malignant transformation, underscoring the importance of complete surgical excision and histopathological examination [60, 61, 62, 63].

Mucous Retention Cysts

Congenital mucous retention cysts are benign cystic lesions that develop due to the accumulation of mucus within glandular ducts. These cysts are typically present at birth and result from an obstruction or dilatation of the ducts, preventing normal mucus drainage. The prevalence rate of mucoceles is reported to be 2.4 cases per 1000 persons, with the highest percentage (70%) observed in individuals aged between 3 and 20 years old [64]. They can occur in various anatomical sites, including the oral cavity [65]. Clinically, mucous retention cysts manifest as painless, fluid-filled swellings that are often translucent and palpable or may reside beneath the mucosal surface. While they are generally asymptomatic, larger cysts may cause discomfort or interfere with oral functions [66]. Diagnosis is confirmed through clinical examination and imaging studies, by means of ultrasound or MRI, to assess the cyst's size and location [67]. Treatment for mucous retention cysts is usually conservative, with surgical intervention reserved for symptomatic cases [64].

Malignant Tumors

Malignant tumors of the oral cavity are exceptionally uncommon among pediatric patients, constituting less than 10% of all pediatric tumor cases [12]. The limited prevalence of this kind of tumor in children undercut the availability of reliable data concerning incidence, recommended treatments, and prognosis. The two primary categories of malignant tumors are teratomas and rhabdomyosarcoma. Due to the rarity of these tumors, comprehensive research and well-documented cases are essential for strengthening our understanding about their epidemiology, optimal therapeutic approaches, and long-term outcomes.

Teratomas

Teratomas are characterized by tissues derived from multiple embryonic germ layers and can manifest in various anatomical locations throughout the body, spanning from intracranial sites to the sacroiliac regions [1]. Overall, teratomas is a rare entity, constituting approximately 1 in 13,000 births worldwide [68]. Oral teratomas account for a relatively small proportion, ranging from 2% to 9% of all teratomas, with the majority originating from the palate [69]. Notably, rare tongue teratomas may arise due to the abnormal segregation of pluripotent germ cells in Rathke's pouch during embryonic gonadal development [70]. A literature review indicates that the incidence of lingual teratomas is comparable between males and females [71]. It has been suggested that maturity of lingual teratomas is not indicate their malignancy.

Head and neck teratomas are histologically categorized into four types [72]: (1) dermoid, characterized by epidermal and mesodermal components; (2) teratoid, consisting of poorly differentiated ectoderm, mesoderm, and endoderm; (3) true teratoma, encompassing all three germ cell layers with identifiable early organ differentiation and a somatic-type malignancy; and (4) epicanthus, highly differentiated with identifiable organs or extremities but associated with a high mortality rate. The histological classification provides valuable insights into the diverse nature of head and neck teratomas, guiding both diagnostic and therapeutic considerations for these congenital lesions [73]. The prognosis of oral teratomas is dictated by the tumor's location and size. Oral teratomas have rather challenging prognosis, primarily due to the impact of the mass on surrounding structures, particularly the fetal airway and structures associated with swallowing mechanisms. The development of oral teratomas, including trisomy 13, is found to be associated with chromosomal abnormalities, and the tumors have been detected in patients with genetic syndromes, such as Aicardi syndrome and Pierre–Robin syndrome. Thus, prenatal genetic testing, particularly chromosomal karyotyping, is recommended to identify any potential chromosomal abnormalities early in the gestational period [50]. The integration of genetic testing in the prenatal evaluation process contributes to a more thorough understanding of the underlying factors associated with these congenital tumors. While most tongue teratomas are typically benign, the likelihood of malignancy tends to rise with age.

The gold-standard management approach for teratomas is early and complete tumor excision [74]. A literature review indicates that most lesions can be successfully and completely resected, resulting in a low recurrence rate [75]. Oral teratomas present on the ultrasound as a cystic-solid mass, which often leads to tongue displacement and potential protrusion from the oral cavity. Color Doppler may reveal blood vessels extending from the fetal palate or tongue into the mass. The presence of calcification aids in the identification of the disease. MRI, though less likely to detect calcification, offers the advantage of delineating a large mass's origin and anatomical relationships, serving as a valuable complement to ultrasound [76]. Nevertheless, the definitive diagnosis of teratoma relies on histopathologic classification. While making a differential diagnosis for oral teratoma, other conditions such as lymphatic malformations, cystic hygroma, hemangiomas, granulosa cell tumors, lingual thyroids, and thyroglossal duct cysts should be considered [50].

Rhabdomyosarcoma

Rhabdomyosarcoma (RMS)—the predominant soft tissue sarcoma in childhood—comprises approximately 4.5% of all pediatric cancer cases. RMS is known for its aggressiveness, invading surrounding tissues [76]. While this type of cancer can originate from various tissues throughout the body, its most prevalent presentation occurs in the head and neck region, constituting approximately 35% of cases [77]. Despite its common occurrence in the head and neck, the tongue represents an atypical site for the development of RMS, with only a few documented cases in the existing medical literature [78, 79, 80, 81]. The rarity of this tumor in the specific anatomical location of the tongue highlights the distinctive nature of its presentation in pediatric patients.

Treatment strategies for RMS are often extrapolated from the general management of pediatric RMS. The primary modalities include a combination of surgery, chemotherapy, and sometimes radiation therapy. Surgical resection is conducted to achieve complete tumor removal while preserving as much healthy tissue and function as possible [82]. Chemotherapy, often consisting of multi-agent regimens, plays a crucial role in reducing tumor size and addressing potential microscopic spread. The inclusion of chemotherapy in the treatment regimen for congenital RMS of the tongue is rationalized by the presumption that these patients may harbor micro-metastases at the time of diagnosis [83]. Radiation therapy may be considered, especially in cases where complete surgical resection is challenging or targeting residual disease. The choice and sequence of these modalities depend on factors such as tumor size, location, and histological subtype [84, 85, 86]. Given the potential long-term effects on speech and swallowing, careful consideration should be given to maintaining optimal quality of life during and after treatment.

Close follow-up is essential to monitor recurrence and manage potential side effects of therapy. RMS typically exhibits distinctive imaging characteristics. On T1-weighted images, RMS demonstrates hypo-to-isointensity compared to muscle, whereas on T2-weighted images, it tends to appear hyperintense. Additionally, avid enhancement is observed on T1 contrasted images, although the enhancement pattern may be heterogeneous.

Discussion

This comprehensive review provides a detailed overview of the diverse spectrum of congenital tongue tumors, encompassing their clinical presentation, radiologic features, and contemporary treatment approaches. Our literature analysis of various studies and case reports contributes to the consolidation of the current knowledge [1, 2, 3, 4], offering a valuable resource for healthcare professionals involved in the diagnosis and management of these rare pediatric lesions. One key insight that emerges from this review is the critical role of advanced imaging techniques in the accurate diagnosis and characterization of congenital tongue tumors. Modalities such as ultrasound, MRI, and CT, along with specialized techniques like diffusion-weighted imaging and perfusion studies [6, 7, 8, 9, 10, 11, 12], have significantly enhanced our ability to differentiate between benign and malignant lesions, as well as to delineate the extent of tumor involvement.

The heterogeneity of congenital tongue tumors reflects the importance of adopting a multidisciplinary approach, with close collaboration between pediatric otorhinolaryngologists, radiologists, and oncologists [5], to ensure optimal patient care. It is important to acknowledge that the decision-making process behind choosing one treatment approach over another is complex and multifaceted, involving consideration of factors such as patient age, tumor size, location, and potential impact on vital structures [20], which are likely to influence treatment decisions. In addition, a more careful consideration of factors such as the risks and benefits of each treatment modality [13, 14, 15, 16, 17, 18, 19] is necessary to gain valuable clinical insights for managing these challenging cases. For instance, surgical excision remains the cornerstone of treatment for many congenital tongue tumors, particularly localized, well-circumscribed lesions [13], but the potential risks of surgery, such as bleeding, infection, and scarring, must be weighed against the benefits of complete tumor removal and the prevention of recurrence [14]. Similarly, embolization and sclerotherapy have shown promise in the treatment of vascular malformations [17, 18], but these procedures can be risky too, causing inadvertent damage to adjacent structures or resulting in incomplete tumor obliteration. Furthermore, treatment modality may be selected based on the consideration of the patient's age, overall health status, and the potential impact of the tumor on vital functions such as breathing, swallowing, and speech [20]. In some cases, a more conservative approach, such as observation or targeted medical therapy, may be appropriate, particularly for smaller, asymptomatic lesions or in cases where the risks of invasive procedures outweigh the potential benefits [15, 16].

The limitations of the studies and the potential for bias in case reports cited in this review should all be acknowledged while interpreting their findings. Many of the studies included in this review are case reports or small case series [2, 4, 11], which, while valuable for describing rare conditions, have inherent limitations. Case reports may be subject to publication bias, as cases with unusual presentations or successful outcomes are more likely to be reported than those with typical features or suboptimal results [31]. Additionally, case reports are not a good source of literature for comparing the effectiveness of specific interventions or the natural history of the condition due to the lack of a control group [32]. Furthermore, the retrospective nature of many of the studies included in this review [1, 3, 6] may introduce bias due to incomplete data collection, inconsistent reporting, or loss to follow-up. These limitations can af-

fect the reliability and generalizability of the findings, rendering the formulation of evidence-based guidelines for the diagnosis and management of congenital tongue tumors extremely challenging.

To address these limitations, future research should focus on conducting larger, prospective studies with standardized data collection and long-term follow-up. Multicenter collaborations and the establishment of patient registries [33] can help to overcome the challenges associated with studying rare conditions and provide more robust evidence to guide clinical decision-making. Moreover, there is a need for high-quality, controlled studies comparing the effectiveness of different treatment modalities for congenital tongue tumors. While the current literature provides valuable insights into the range of available options, the lack of direct comparisons makes it difficult to determine the optimal approach for a given patient [34]. Future research should aim to evaluate the outcomes, complications, and long-term effects of various interventions, taking into account factors such as tumor type, size, location, and age of patients.

Another crucial aspect that warrants further exploration is the long-term outcomes and quality of life implications for children with congenital tongue tumors. Although data in the literature report potential functional impairment and underscore the need for a multidisciplinary approach to management [36, 37, 38, 39, 40], the psychosocial impact of these conditions and the importance of long-term follow-up and support must be investigated to provide a more comprehensive understanding of the challenges faced by these patients and their families. Congenital tongue tumors may have a profound impact on a child's development, affecting not only their physical health but also their emotional well-being and social interactions. Depending on the size and location of the tumor, children may experience difficulties with feeding, speech, and swallowing [36, 37], which can lead to nutritional deficiencies, delayed language development, and social stigma. These challenges can persist even after the tumor has been treated, requiring ongoing support from a multidisciplinary team of healthcare professionals, including speech therapists, nutritionists, and psychologists [38]. Moreover, the psychosocial impact of congenital tongue tumors may extend to the family members of the affected children. Parents may experience significant emotional distress, financial burden, and social isolation as they navigate the complex healthcare system and adapt to their child's unique needs [39]. Siblings may also be affected, as they may experience feelings of neglect or resentment due to the increased attention and resources directed toward the affected children [40]. To address these challenges, long-term follow-up and support are essential. Regular monitoring of the pediatric patient's growth, development, and functional abilities can help identify and address any ongoing issues related to the tumor or its treatment [41]. Psychosocial support, including counseling and peer support groups, can help the affected children and their families cope with the emotional and social challenges associated with these conditions [42]. Educational interventions, such as workshops and written materials, can also help families understand the condition better and develop strategies for managing its impact on daily life [43]. Furthermore, research into the long-term outcomes and quality of life of children with congenital tongue tumors is needed to develop evidence-based guidelines for follow-up care and support. Longitudinal studies that assess the physical, emotional, and social well-being of these children over time are also warranted to shed light on the long-term impact of these conditions and to identify factors that contribute to positive outcomes [44]. Qualitative research, such as interviews and focus groups with affected children and their families, can also provide a more nuanced understanding of their experiences and needs [45].

Given the rarity of these tumors [23, 24, 25, 26], international collaborations and the establishment of multi-center registries should be fostered to advance research that contribute to enhancing our understanding of these conditions. Through pooling data and expertise across institutions, we are armed with better tools to identify patterns, refine diagnostic criteria, and develop evidence-based treatment protocols. Such collaborations would also facilitate the sharing of best practices and the development of standardized guidelines for the management of congenital tongue tumors.

Conclusions

Congenital tongue tumors constitute a fascinating yet rare facet of pediatric pathology, encompassing a diverse spectrum of hemangiomas, vascular malformations, dermoid cysts, teratomas, and rhabdomyosarcomas. Despite their rarity, these tumors present unique clinical behavior, diagnostic features, and therapeutic intervention challenges. The clinical presentation of congenital tongue tumors varies widely, spanning from visible swelling to more severe manifestations like dysphagia and respiratory distress. Accurate diagnosis is contingent upon a combination of clinical evaluation and imaging examinations through ultrasound, MRI, and CT. Fortunately, each tumor type exhibits distinct radiologic features, aiding in the differentiation and characterization of these congenital lesions. Additionally, the integration of genetic testing in prenatal evaluation contributes to a more thorough understanding of the underlying factors associated with these tumors.

Treatment strategies for congenital tongue lesions are multifaceted and often involve a multidisciplinary approach. Surgical excision stands as the mainstay of congenital tongue tumors management. Many different treatment modalities for this kind of lesion are available, but they are selected after considering tumor type, size, and potential implications for vital structures. While most congeni-

tal tongue lesions are benign, some variants are malignant, like teratomas and RMS, underscoring the need for early and complete excision.

Advanced imaging techniques and a multidisciplinary approach involving pediatric surgeons, radiologists, and oncologists are key to optimizing patient care and improving outcomes in this challenging field. The prognosis of these tumors is influenced by factors such as tumor's location, size, and impact on surrounding structures. Close follow-up is crucial for monitoring recurrence and managing potential side effects of therapy.

This comprehensive review highlights the importance of a tailored approach to the diagnosis and management of congenital tongue tumors, taking into account the distinctive clinical, radiological, and histopathological features of each entity. The future management of congenital tongue tumors is likely to be shaped by emerging treatment modalities, such as targeted therapy and minimally invasive techniques, offering more personalized and effective care for the affected patients. Consolidating current knowledge into the broader landscape of pediatric otolaryngology could further understand these rare conditions and promote advances in the diagnosis and management of congenital tongue tumors. Continued research and documentation are imperative to enhance our understanding of these rare pediatric tumors' epidemiology, optimal therapeutic approaches, and long-term outcomes.

Availability of Data and Materials

Not applicable.

Author Contributions

AM: conception and design, interpretation of data; LLV: acquisition of data, analysis; SC: analysis, interpretation of data; PP: acquisition of data, conception and design; AM, LLV and SC: writing the draft; ML, SL: conceptualization; GI: formal analysis. All authors revised the manuscript critically for important intellectual content. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

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Conflict of Interest

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