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Ancient schwannoma mimicking a carotid body tumor: a case report and review of the literature

Ancient Schwannoma is a type of peripheral neurogenic tumor formed by the Schwann cells presenting mainly as a benign and asymptomatic lesion. The neurilemmoma tumor appears in different sites and, in cases of cervical location, can mimic a carotid body tumor. Herein we describe a clinical case of a 51-year-old woman with latero-cervical swelling. A contrast-enhanced computed tomography scan revealed a classical wineglass image mimicking a carotid body tumor. During surgery the tumor appeared connected to the cervical sympathetic trunk without carotid involvement. The histological exam confirmed the nature of the mass which consisted of an ancient schwannoma. A subsequent systematic review of the literature on ancient schwannoma incidence and treatment confirms it being a benign and rare lesion primarily treatable with open surgery.

KEY WORDS: Ancient Schwannoma, Carotid body tumor, Carotid artery, Vascular surgery

Introduction

Schwannoma is a rare tumor derived from the peripheral nervous system stemming from Schwann cells; a type of cell that wraps itself around peripheral nerves and provides protection and support. Schwannoma is almost always benign, but it can rarely present as a malignant degeneration. It can occur sporadically or within the context of a family syndrome known as neurofibromatosis [1]. This type of tumor is slow growing and, in most cases, is asymptomatic. When symptomatic it can be detected by radiating pain, muscle weakness, tingling, a "*pins and needles*" sensation, or numbness but symptoms reflect the involvement of the peripheral nerve affected [2]. Approximately 25-40% of all reported schwannomas occur in the head and neck [3-6] with most of these affecting the eighth nerve. Schwannoma of the cervical sympathetic chain is a rare type appearing as an asymptomatic and encapsulated solitary neck mass located in the posterior cervical region at paravertebral level. Definitive pre-operative diagnosis may be difficult and usually requires multiple imaging tests while only the histological exam can reveal the tumor's real nature.

Clinical case

Our patient was a 51-year-old woman, a native of the Eastern Europe, a smoker, affected by hypertension. During the clinical exam, a right latero-cervical hard and confined swelling was noted without any other symptoms (Fig. 1). A duplex scan examination suggested a carotid body tumor. A contrast-enhanced computed

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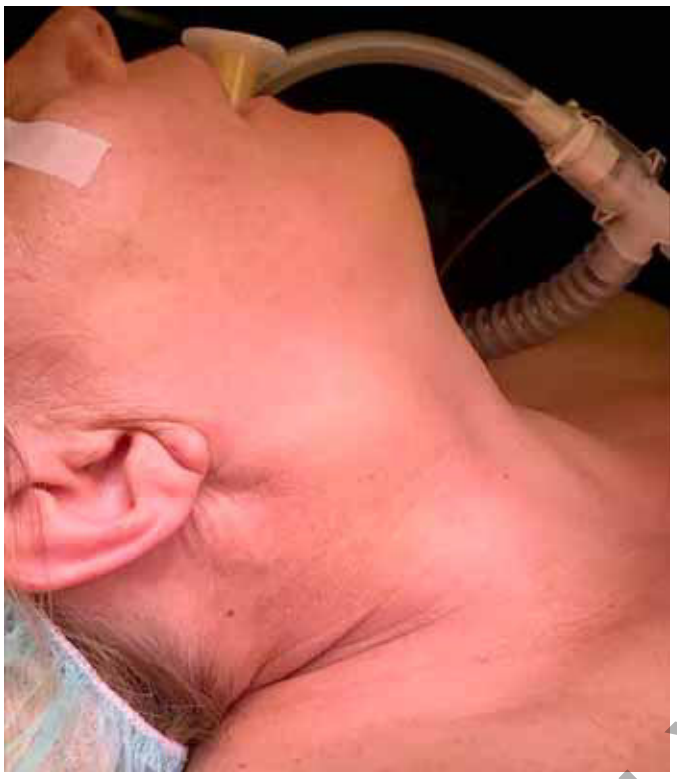

Fig. 1: Photo of the patient: it's evident the right latero-cervical mass.

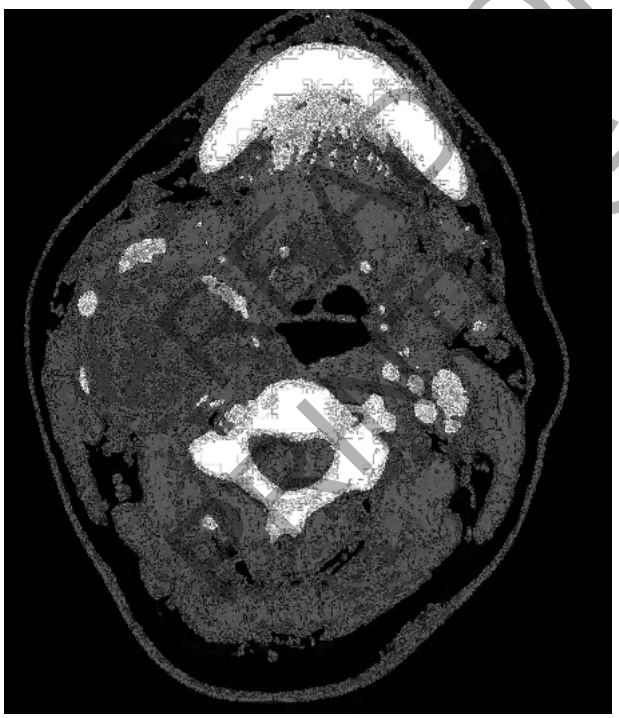

Fig. 2: Contrast-enhanced CT scan.

tomography (CT) scan of the neck revealed a solid, well-defined latero-cervical mass measuring approximately 45 mm x 25 mm x 2 mm, appearing as a carotid body tumor with a classical wineglass imagine (Fig. 2). A preoperative arteriography and subsequent embolization was planned in order to reduce perioperative bleeding during the surgical procedure, however it was impossible to cannulate the glomic artery due to the low vascularization of the mass (Fig. 3). One day after the CT scan, surgical intervention was performed with latero-cervical access to the carotid artery making an incision along the anterior border of the sternocleidomastoid muscle. During isolation of the mass it was possible to note that the carotid artery was not involved in the lesion (Figs. 4a-c) and while dissecting from the surrounding structures it became clear that the mass was independent from the vagus and hypoglossal nerves. Proceeding superiorly and deeply following the glossopharyngeal nerve, we discovered that the mass was not involved but rather appeared to be connected to the cervical sympathetic trunk. The tumor enucleation was performed without sacrificing the nerve fibers which were pushed aside and the capsule cut open, allowing for the resection of the schwannoma. A histopathological examination of the anatomic spe-

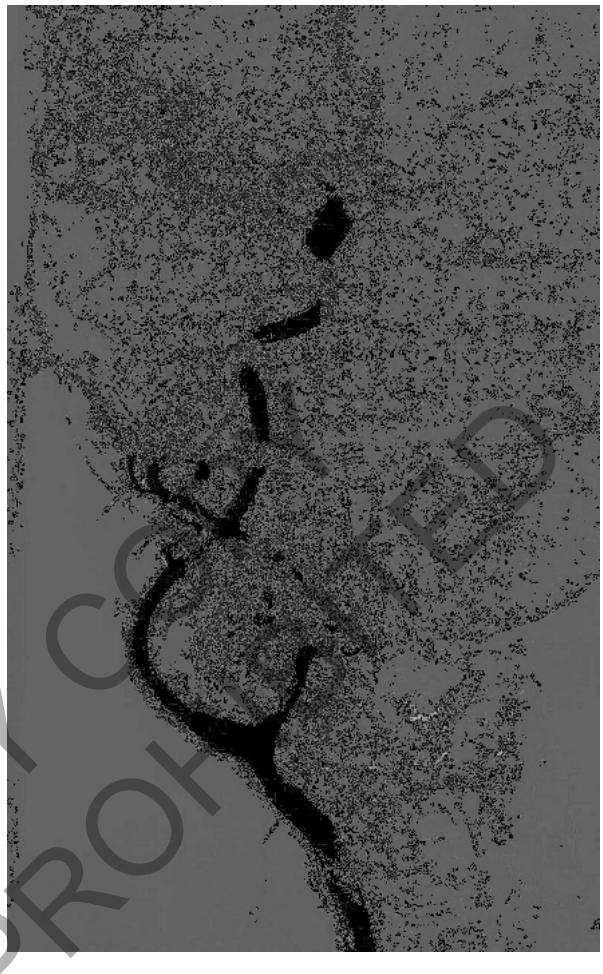

Fig. 3: Preoperative digital subtraction angiography: a classical wineglass image is shown mimicking the presence of a carotid body tumor.

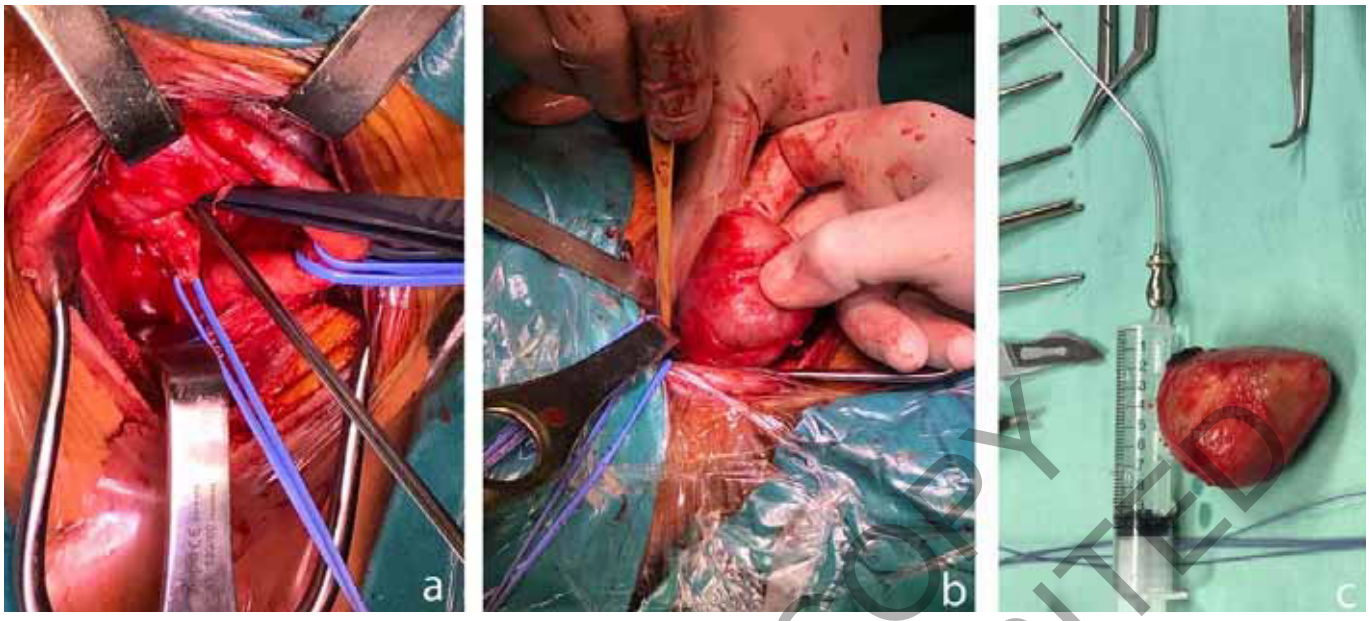

Fig. 4: Intraoperative pictures: during carotid bifurcation dissection (a); mass resection (b); and final view of the resected mass (c).

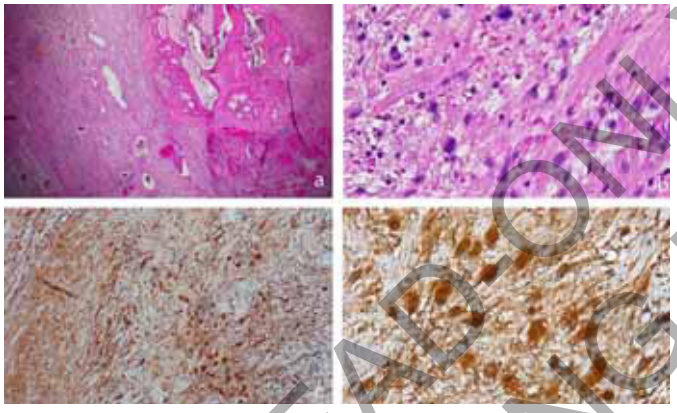

Fig. 5: Focally palisading nuclei of spindle cells represented by a Verocay body, marked myxoid degeneration and hyalinisation are typical histological findings of this tumour. [Haematoxylin and Eosin 4X(a) and 20X(b)]. The immunohistochemical exam documents the positivity for the protein S 100, diffusely positive for this tumour (c-d).

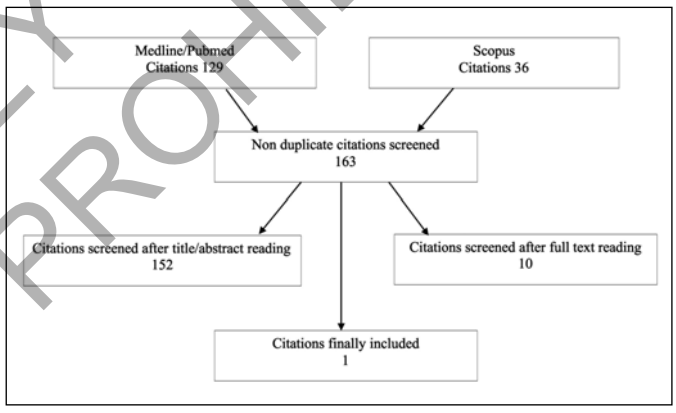

Fig. 6: Flowchart of systematic review.

cimen established a diagnosis of *ancient schwannoma* (Figs. 5 a-d). The postoperative course was smooth without any complications reported and the patient was discharged three days after surgery. A clinical and duplex-scan follow-up was performed at 3, 6 and 12 months after the intervention. The patient was in satisfactory clinical condition and has not shown any post-operative complications, neurological disorders and recurrences of pathology after 20 months post-operatively.

Discussion

Five different types of schwannoma are described in the literature: common, plexiform, epithelioid, cellular and ancient type[4,7]. The terminology "ancient" schwannoma was first suggested by Ackerman and Taylor in a review of 48 thorax neurogenic tumors, 10 of which appeared similar to those of typical neurilemmomas but differing in that significant tumor portions contained only a few cells within hyalinized matrices [8]. These same features were found in long time existing schwannomas and for this reason the term "ancient schwannoma" was proposed to indicate the protracted period and degenerative changes associated with the appearance of such lesions particularly: hyperchromatic areas and nuclear atypia with variable presence of myxoid, fibrillary background and cystic degenerative changes [4]. Ancient type schwannomas occur within a wide age range between 20-50 years and is predominant in females [4,9]. The majority of

patients are asymptomatic and the presence of Horner's syndrome before resection is possible but rare[4]. The "lyre sign" of CT, with displacement of the internal and external carotid artery, can occur in schwannomas of the cervical sympathetic chain although displacement of the carotid artery and jugular vein together is typical and more frequent [4]. Clinical presentation is usually an isolated asymptomatic lateral-cervical mass [10]. Imaging by CT or Magnetic Resonance Imaging (MRI) is mandatory in order to manage and is essential to discover tumor size, extension, and vascular relation [10]. MRI contrast is able to identify schwannomas pre-surgically and also provides information about the origin of the nerve enabling adequate patient counseling before being subject to surgical excision [4] however proper identification of the nerve is often impossible until the operation which is the treatment of choice for this tumor [3,11]. Two different surgical options for schwannoma treatment are described in our recent literature review: the first is the gross total resection that completely excises the tumor with or without nerve sacrifice, and the second is subtotal resection that is a nerve-sparing technique which leaves the tumor capsule behind [1]. After surgery only the histological exam can establish the type of tumor existing while the protein S 100 is diffusely positive in the immunohistochemical exam in the ancient subtype[4,12].

As known, a fibrous capsule separates the nervous fibers from the schwannoma, unlike neurofibroma where the nerve is an integral part of the tumor [11] and therefore it is generally possible to enucleate the tumor without causing meaningful nervous lesions**.** If the sacrifice of the nerve is necessary, Horner's syndrome is the most frequent post-operative complication. Even if clinical and instrumental diagnosis of schwannoma is formulated, surgical treatment is often necessary due to the impossibility of discriminating the nature of the lesion prior to surgical intervention [11]. Also, in their progressive growth stage, schwannoma have the tendency to displace, compress and damage the surrounding structures thus making the intervention more difficult and burdened by higher possible complication rate. Being radioresistant as well, surgery is the best choice. The most important, though rare, risk related to surgery is nerve injury which occurs most frequently in schwannoma type due to the close relationship these tumors have to the nerve's route. Nerve injuries may be direct due to trauma or cut, or indirect due to compression or stretching [11]. Reported incidence of neurological deficits after schwannoma resection varies greatly between 1.5 and 80 percent. A significantly high percentage of complications occur in short-term observations and the majority of neurologic complications are transient [13]. It is useful to differentiate iatrogenic nervous paralysis from post-surgical stupor paresis which is a transient condition caused by inflammatory phenomena. Finally, in accordance with reported literature, we believe that the risk of exacerbation of compression neuropathy caused by gradual tumor growth justifies surgical intervention [13].

A literature review on ancient schwannoma of the sympathetic chain was carried out in the PubMed and Scopus databases. The search was conducted by two authors (R.G. and D.T.) with string: (ancient schwannoma OR neck schwannomas OR cervical sympathetic chain tumor OR cervical tumor) AND (carotid paraganglioma OR carotid surgery OR carotid tumor) AND (resembling OR mimicking OR differential diagnosis OR masquerading)". Results were processed by a PRISMA statement. Only literature from 1952 to 2018 published in the English language was considered. 129 articles from PubMed and 36 from Scopus, of which 163 were non duplicate citations, were screened. One-hundred-fifty-two (152) after title/abstract reading and 10 after full text reading were excluded. Finally, 1 citation regarding sympathetic chain ancient schwannoma during carotid surgery was included as it proved that it was possible to find a sympathetic chain schwannoma mimicking a carotid body tumor but that the histological ancient type is a very rare entity. The study flowchart selection is reported in Fig. 6 and includes all case reports. In most investigated scientific articles, the histological examination of the schwannoma did not specify histological class of membership. Indeed, many studies examined reported clinical cases that generically concern schwannoma of the sympathetic chain and not specifically the ancient type which could explain the reason why our research led to a single case. We therefore believe that, although ancient schwannoma of sympathetic chains is a rare lesion, it is necessary to further study this tumor along with a detailed histological examination of the specimen as reported in the scientific articles so as to know its real frequency. In conclusion ancient schwannoma of the cervical sympathetic chain is a rare and often asymptomatic entity. Contrast enhanced CT scan and MRI are indispensable diagnostic tools but only the histological examination can define tumor type. Lastly, the published literature, as noted, is in line with our experience in treatment of schwannoma of the sympathetic chain confirming that surgery allows for both specific diagnosis and final therapy.

Riassunto

L'*ancient schwannoma* è un tumore che origina dalle cellule di Schwann presenti nella guaina dei nervi periferici. Si presenta solitamente come lesione benigna ed asintomatica. Questo tipo di tumore può essere riscontrato in differenti sedi, e nei casi in cui si localizza a livello cervicale, tale lesione può apparire come un tumore del glomo carotideo. Descriviamo in questo articolo un caso clinico di una donna di 51 anni con una tumefazione latero-cervicale. La TC preoperatoria mostrava la classica immagine "wineglass", a calice, tipica del tumore del

glomo carotideo con divaricazione della biforcazione carotidea. Durante l'intervento chirurgico la lesione sembrava adesa alla catena cervicale del simpatico senza coinvolgimento dei vasi carotidei. L'esame istologico ha confermato che tale massa era uno schwannoma del tipo "ancient". Infine abbiamo effettuato una revisione sistematica della letteratura sull'incidenza e il trattamento dell'ancient schwannoma, confermando quest'ultima come una lesione benigna e rara, trattabile preferibilmente con la chirurgia aperta.

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