Orbital lesions, an interdisciplinary pathology. The experience of the maxillo-facial surgeons



Ann Ital Chir, 2022 93, 3: 271-279 pii: S0003469X22036107 Online ahead of print 2022 - Jan. 10 *free reading*: www.annitalchir.com

Valentina Terenzi*/**, Andrea Cassoni*/***, Resi Pucci*, Marco Marenco****, Maria Teresa Fadda*/***, Ingrid Raponi*, Marco Della Monaca*/***, Valentino Valentini*/***

*Department of Oral and Maxillofacial Sciences. Sapienza University of Rome, Italy

**ENT Unit, Ospedale "Santa Maria Goretti" Azienda USL Latina, Italy

***Oncological and Reconstructive Maxillo-Facial Surgery Unit, Policlinico "Umberto I" di Roma, Department of Sense Organs.

"Sapienza" University of Rome, Rome, Italy

****Department of Sense Organs "Sapienza" University of Rome, Rome, Italy

Orbital lesions, an interdisciplinary pathology. The experience of the maxillo-facial surgeons

AIM: The main focus of this paper was to describe our experience in the management of primary lesions involving the orbit, analyzing signs and symptoms and illustrating surgical management.

MATERIAL OF STUDY: We describe our experience managing 62 consecutive patients diagnosed with orbital tumors confined to the orbital content (not involving skin or bone, with no intracranial invasion) and treated between January 2013 and December 2016. Demographic characteristics, symptoms, clinical findings, histological types and approaches have been recorded.

RESULTS: We found that the most common clinical manifestations were exophthalmos/proptosis and ocular movement impairment; the most common histological types were vascular malformation and ocular adnexal lymphomas.

DISCUSSION: Primary neoplasms involving orbital contents include a broad spectrum of pathologies difficult to manage without a firm diagnosis, usually histological. Different surgical accesses are described in order to perform incisional biopsy or resection of the mass.

CONSCLUSION: In every case, accurate surgical planning is mandatory, in order to prevent functional and/or aesthetic complications.

KEY WORDS: Blepharoplasty incision, Intraorbital neoplasia, Lateral orbital tumor, Orbitotomy, Surgical treatment, Transconjunctival approach,

Introduction

The orbit can be considered as an "interdisciplinary" region, as it is an area of interest for many specialists, such as ophthalmologists, ENT (ear, nose, and throat) doctors, endocrinologists, neurosurgeons, plastic and maxillofacial surgeons. Primary neoplasms involving

orbital content include a broad spectrum of pathologies; in literature, their prevalence varies according to age and region of study ¹. They can be divided into benign lesions as vascular malformations and inflammatory deseases and malignant neoplasms involving the orbit as ocular adnexal lymphomas, or metastases ^{2,3,4}. Furthermore, the orbit can be affected by malignant or benign lesions originating from nearby structures ^{5,6}. We can distinguish between intraconal and extraconal diseases: the first ones are usually benign, slow-growing masses, while the others are rarer and usually involve the orbit secondarily⁷. In this paper, we retrospectively describe our experience, from 2013 to 2016, in the management of primary lesions involving the orbit, analyzing signs and symptoms and illustrating surgical management.

Pervenuto in Redazione Luglio 2021. Accettato per la pubblicazione Settembre 2021

Correspondence to: Resi Pucci, Department of Oral and Maxillofacial Sciences, Sapienza University of Rome, Via Caserta 6, 00161 Rome, Italy (e-mai: lresi.pucci@gmail.com)

Materials and Methods

We retrospectively reviewed the data of adult patients with a diagnosis of orbital tumor who presented at our center from January 2013 to December 2016 (minimum 3 years of follow-up). Inclusion criteria were: 1) lesion limited to the orbital content, not involving skin or bone, without intracranial invasion; 2) radiological preoperative imaging (CT and/or MRI); and 3) histological diagnosis. Data on patients' characteristics, symptoms, clinical findings, histological type, site of the lesion, and imaging were recorded in each case.

Results

Sixty-two adult patients were identified (28 males – 41.16% – and 34 females – 54.83%) respecting inclusion criteria, with a median age of 52 years (range: 18-85). Preoperative imaging (CT or MRI) showed that 56.45% of the lesions were located in the right orbit; no bilateral lesions were found. Main data included: characteristics, signs and symptoms (proptosis, pain, epiphora, motility impairment), tumor localization (intraconal or extraconal), surgical management (incisional biopsy or complete surgical removal), and surgical approach (transconjunctival approach, upper eyelid skin crease incision with or without orbitotomy, lateral orbitotomy with coronal scalp flap approach. All the data collected are displayed in Table I.

Signs and Symptoms

42 patients (67.7%) presented exophthalmos/proptosis in both benign and malignant neoplasms (29 patients – 69% – with benign lesions and 13 patients – 31% – with malignancy). Ocular motility impairment was present in most cases of intraorbital metastasis (75%), in patients with vascular malformation (46%) and orbital pseudotumor (37.5%), while it was rarely observed in other cases. Pain was present in two patients affected by metastasis. All clinical symptoms are shown in Table I.

SURGICAL MANAGEMENT

Total surgical removal of the lesion was performed on 36 patients. The definitive histological diagnosis confirmed 15 vascular malformations, 2 idiopathic orbital inflammations (chronic dacryoadenitis), 8 benign neural tumors, 8 dermoid cysts, 2 benign tumors of the lacrimal glands, and 1 metastasis. The biopsy was performed on 26 patients: it confirmed the diagnosis of malignancy in 16 patients (13 lymphomas of the annexes and 3 metastases) and the diagnosis of inflammatory lesion in 10 patients. In all patients, the selected surgical approach allowed to obtain a histological diagnosis, and no surgical complications/sequelae were identified in the follow-up period.

SURGICAL APPROACHES

Transconjunctival approach (19 patients, 30.6%)

Indications: it can be used to gain access to the orbit for incisional biopsy or excision of orbital tumors on the inferior wall ⁸.

Technique: lower eyelid is infiltrated with 1% lidocaine with epinephrine 1:100,000; a 5–0 silk suture is passed through the Gray line so that the eyelid is everted over a Desmarres retractor and the conjunctival incision is made about 3 mm inferior to the tarsal border. Orbital tissue is retracted using a malleable retractor, to explore the intraorbital content. The surgeon must avoid exerting too much pressure on the eyeball in order to limit the risk for complications, even severe.

Blepharoplasty incision with or without lateral orbitotomy (30 patients, 48.3%)

Indications: it can be used to gain access to the superior and lateral orbit (i.e., lacrimal gland) ⁹.

Technique: upper eyelid is infiltrated with 1% lidocaine with epinephrine 1:100,000; an incision is marked on a naturally occurring skin line in the supratarsal fold, parallel to the superior palpebral sulcus. The incision can be extended into the crow's feet of the lateral orbital skin. After incision, a skin-muscle flap is developed superficial to the orbital septum/elevator aponeurosis complex and retracted to expose the periosteal surface. Orbitotomy technique: once that the periosteal surface is exposed, an incision is made over the superolateral orbital rim. The dissection proceeds on a subperiosteal plane, in order to expose the lateral orbital wall. After pre-plating, the osteotomy is performed using piezosurgery. The bone flap can be removed or dislocated and then fixed using titanium plates.

In the upper eyelid skin crease incision group, 9 patients were treated with upper eyelid blepharoplasty incision with orbitotomy (9 patients, 14.5%).

Coronal flap with lateral orbitotomy (13 patients, 20.9%) Indications: it can be used to gain access to large lesions in the superior and lateral orbit and in the orbital apex¹⁰. Recently, this approach is being used less frequently, also because of the progress in the anatomical knowledge and surgical techniques involving this area ¹⁰.

Technique: after infiltration with 1% lidocaine with epinephrine 1:100,000, a coronal incision is performed, and a scalp flap is reflected to expose the orbital rim. Dissection is performed on the subperiosteal plane, so that it possible to perform the osteotomy in the way described previously. The flap can be removed or dislocated and then fixed using titanium miniplates after tumor biopsy or removal.

Table I - The sample characteristics (median age and SD: standard deviation, male and female expressed in %); clinical presentation (proptosis/exophthalmos, pain, epiphora, and OMI: ocular motility impairment); surgical management (incisional biopsy or complete surgical removal); surgical approach (transconjunctival, upper eyelid skin crease, and lateral orbitotomy with coronal scalp flap); tumor localization (intraconal or extraconal).

	Characteristic	Clinical Presentation	Surgical treatment	Surgical Approach	Side	Tot. Pt 62
VASCULAR MALFORMATION	Age (years)	Esophtalmuos/proptosis 53.3%(8)	Surgical biopsy -	Transconjunctival 40%(6)	Intraconal 80%(12)	15 24.3%
Orbital Venous Malformation	Median 47.20 S.D. +/-17.14 Gender:	Pain 13%(2) Epiphora O.M.I. 46.6%(7)	Complete surgical removal 100%	Blepharoplasty incision 40% (6) Coronal/lateral orbitotomy* 20% (3)	Extraconal 20%(3)	
OCULAR ADNEXAL LYMPHOMA	Age (years) Median 71 S.D. +/- 10.36 Gender: Male 30.77% Female 69.23%	Esophtalmuos/proptosis 69.2% (9) Pain 15.38% (2) Epiphora 69.2% (9) O.M.I. 15.38% (2)	Surgical biopsy 100% Complete surgical removal	Transconjunctival 28.5% (5) Blepharoplasty incision 61.5%% (8) Coronal/lateral orbitotomy*	Intraconal 46.2% (6) Extraconal 53.8% (7)	13 20.9%
IDIOPATHIC	Age (years)	Esophtalmuos/proptosis	Surgical biopsy	Transconjunctival	Intraconal	12
ORBITAL INFLAMMATION . Pseudotumor 66.6% (8) . Chronic dacryoadenitis 16.6% (2)	Median 48.25 S.D. +/-20.49 Gender:	62.5%(5) Pain Epiphora O.M.I. 37.5%(3)	83.3% (10) Complete surgical removal 26.7% (2)	37.5%(7) Blepharoplasty incision 62.5%(5) Coronal/lateral orbitotomy*	63.6% (7) Extraconal 36.4% (5)	19.3%
DERMOID CYST	Age (years)	Esophtalmuos/proptosis	Surgical biopsy	Transconjunctival	Intraconal	8 12.9%
DERMOIDCISI	Median 42.63 S.D. +/-17.98 Gender:	62.5%(5) Pain Epiphora 0.M.I. 37.5%(3)	Complete surgical removal 100%	Blepharoplasty incision 50% (4) Coronal/lateral orbitotomy* 50% (4)	- Extraconal 100%	12.976
NEURAL BENIGN TUMORS	Age (years) Median 46.75 S.D. +/-16.57 Gender:	Esophtalmuos/proptosis 75% (6) Pain 12.5% (1) Epiphora O.M.L 25% (2)	Surgical biopsy - Complete surgical removal 100%	Transconjunctival Blepharoplasty incision 25% (2) Coronal/lateral orbitotomy* 75% (6)	Intraconal 37.5% (3) Extraconal 25% (2) Intracranial origin 37.5% (3)	8 12.9%
METASTASIS	Age (years) Median 58 S.D. +/- 6.06 Gender:	Esophtalmuos/proptosis 100% (4) Pain 50% (2) Epiphora 25% (1) O.M.I. 75% (3)	Surgical biopsy 75%(3) Complete surgical removal 25%(1)	Transconjunctival 25%(1) Blepharoplasty incision 75%(3) Coronal/lateral orbitotomy*	Intraconal 75% (3) Extraconal 25% (1)	4 6.5%
LACRIMAL GLAND TUMORS	Age (years) Median 47.17 S.D. +/-19.68 Gender: Male 50% Female 50 %	Esophtalmuos/proptosis 83.3% (5) Pain Epiphora O.M.I. 17% (1)	Surgical biopsy - Complete surgical removal 100%	Transconjunctival - Blepharoplasty incision 100% Coronal/lateral orbitotomy	Intraconal - Extraconal 100%	2 3.2%

V. Terenzi, et al.

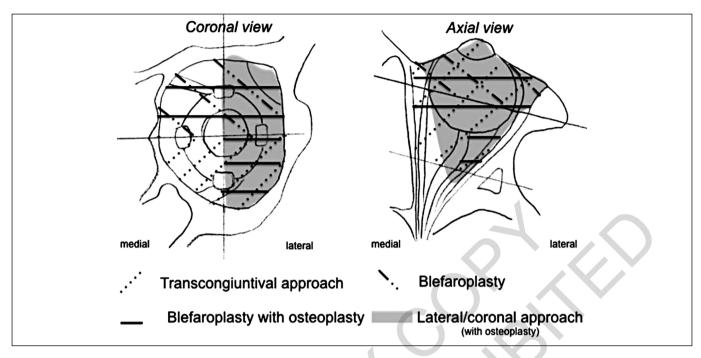


Fig. 1: Surgical field based on approaches described.

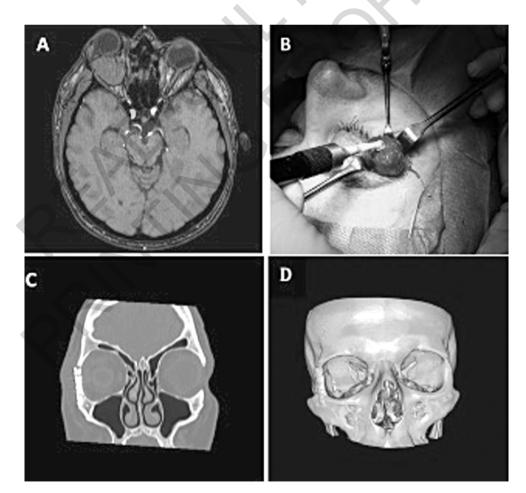


Fig. 2: Clinical case of a patient suffering from orbital hemangioma. A) preoperative MRI image; B) intraoperative detail, removal using the cryostat; C) postoperative CT, axial view; D) postoperative 3D CT scan.

HISTOLOGICAL FINDINGS

Final diagnosis was made based on the surgical pathological results.

Vascular malformations (15 patients, 24%)

According with ISSVA classification¹¹, patients were affected by orbital venous malformation (OVM)¹²; mean age at presentation was 47 years, and females resulted more affected than males (60% vs. 40%); 12 patients presented an intraconal lesion (80%). Clinically, 53% of the sample presented proptosis, 46% motility impairment, and 13% pain. All patients underwent complete surgical resection.

Ocular adnexal lymphoma (13 patients, 21%)

We identified 6 cases of extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma), three of lymphoplasmacytic lymphoma, two cases of follicular center cell lymphoma, and 1 case of diffuse large B-cell lymphoma. Mean age was 71 years (range 19-86 years); 54% of the patients presented an extraconal lesion. A surgical biopsy was performed on all patients, using upper eyelid skin crease incision in 8 cases (61.5%), and a transconjunctival approach on the remaining 5 patients. Clinically, the most frequent symptoms are epiphora and exophthalmos, both present in 69% of patients.

Idiopathic inflammatory pseudotumor (8 patients, 13%) Mean age at presentation was 48 years (range 20-79 years) and males resulted more affected than females (75% vs. 25%). Pseudotumor represents 66.6% of the idiopathic orbital inflammation (IOI) in our sample. Clinically, 62.5% of patients presented proptosis, 37.5% motility impairment, and 12.5% referred pain. All patients underwent a surgical biopsy; transconjunctival and upper eyelid skin crease incision were the two surgical approaches we preferred (37.5% and 62.5%). The lesions were intraconal in 62.5% of the sample, and extraconal in 37.5%.

Dermoid cyst (8 patients, 13%)

Mean age in the group was 42 years (range 31-60 years), No difference in distribution by gender was observed. Clinically, 62.5% of patients presented proptosis, 12.5% presented pain and epiphora. In 37.5% of cases, motility impairment of the eye was present. Complete surgical removal was performed by upper eyelid skin crease incision in four cases, while the remaining patients were treated with a more extended approaches, such as coronal scalp flap and lateral orbitotomy.

Benign neural tumor (8 patients, 13%)

Mean age at presentation was 46 years (range 22-89 years), and females (62.5%) were more affected than males. Four patients presented an optic nerve menin-

gioma, three a plexiform neurofibroma – in one case associated with neurofibromatosis 1 – and one a schwannoma. The lesions were intraconal in 37.5% of patients, extraconal in 25% and the 37.5% had an intracranial origin. Clinically, most patients (75%) presented with proptosis; more rarely, pain (12.5%) or motility impairment of the eye (25%) were observed. Complete surgical removal was performed in all patients, using an extended approach, i.e., lateral orbitotomy with coronal scalp flap, in 75% of cases.

Benign lacrimal gland lesions (6 patients, 10%)

Mean age at presentation was 47 years (range 25-72 years), with equal distribution between the genders. Proptosis was the most common sign (83.3%). Two patients were affected by idiopathic orbital inflammation, two by a chronic dacryoadenitis, and the remaining two patients were affected by benign pleomorphic adenoma. Complete surgical removal was performed on 5 patients (83.4%).

Metastases (4 patients, 6%)

Mean age at presentation was 58 years (range 55-76 years). Females were more affected than males (75% vs 25%), and 75% of patients presented an intraconal lesion. Breast cancer, in women, is the most frequent malignancy metastasizing into the orbit, while one male patient presented metastasis from kidney cancer. Metastatic lesions are the most clinically evident.

Clinically, all patients presented proptosis, associated with motility impairment in 75% of the sample, two patients had pain, and one had epiphora. Complete surgical removal was performed in one case with an upper eyelid skin crease incision with osteoplasty approach. 3 patients underwent surgical biopsy, two via the upper eyelid incision already made, and one via transconjunctival approach.

Discussion and Comments

Orbital tumors are rare, with an incidence of less than 1/100,000, and they usually become apparent only when they reach a diameter of at least 1 cm⁻¹¹. The reported prevalence varies in the literature, according to age, localization, and geographic area of study population ¹. The presence of orbital tumors can lead to exophthalmos/proptosis, impaired ocular motility or alignment, eventually resulting in diplopia, sensory changes including V2 impairment, and visual loss 14. Most frequently, patients present with exophthalmos (67.7%), in most cases monolateral ¹³. Bilateral exophthalmos can be consequent to thyroid-related orbitopathy (in those cases, histological or cytological diagnosis is not required), lymphoma, or orbital inflammation. In all cases, ophthalmological evaluation and MRI are mandatory. Histological (or cytological) diagnosis is fundamental,

except when inflammatory disease (such as pseudotumor) is suspected, since it regresses after corticosteroids. Nevertheless, this behavior can also be observed in lymphomas, so biopsy can be avoided only in those cases in which lymphatic disease can be surely excluded on the basis of clinical-radiological findings ¹³.

Fine-needle aspiration (FNA) can be considered: even if some authors assess that «...in case there are not contraindications it should be performed each time an orbital tumor is diagnosed» 13 , we believe that it has to be excluded if a vascular lesion is suspected, in order to prevent bleeding, and that it is not appropriate in case of suspicion of lymphoma, since the sample is not sufficient to obtain a diagnosis ¹⁵. Even if cytological sampling has the advantage of being less invasive and nearly devoid of complications and dissemination (a particularly dreaded eventuality for pleomorphic adenoma), we have to consider that a large number of samples result "non-diagnostic" and that it is impossible the use of immunostains 15 . In conclusion, we think that, with a larger superficial lesion, FNA may be performed as the first diagnostic step after MRI, if vascular or lymphoproliferative diseases have been excluded. In the first case, it is obviously due to the risk of bleeding, while, in the case of lymphoma, a representative sample is required in order to perform a diagnosis.

Another case in which incisional biopsy has to be avoided is when pleomorphic adenoma is suspected. Once the decision to perform a biopsy is taken, surgical access has to be chosen ^{2,16}. It will depend on localization (intraor extraconal), vascularization, diagnostic suspicion (benign or malignant lesion).

Nevertheless, according to our experience, in most cases this procedure is not completely safe, and it is not well accepted by the patient that, once informed of the risk of obtaining a non-diagnostic sample, prefers surgical biopsy, performed through an upper eyelid skin crease incision. In the suspect of pleomorphic adenoma, complete excision with an intact pseudocapsule is required in order to prevent dissemination and recurrence, possibly preserving the palpebral lobe of the gland, to reduce the incidence of postoperative "dry eye"¹⁷⁻¹⁹. In accordance with what reported above, in our experience, in most cases (83.4%) of lacrimal gland lesions, the biopsy was performed using a "skin crease" approach; in only 2 cases, the pleomorphic adenoma of the lacrimal gland was removed entirely.

Most frequently, the lacrimal gland results affected by inflammatory disease, while lymphoproliferative disorders are the second most common tumor type^{2,20}. Epithelial neoplasms are, in most cases, benign²¹. Other cases in which incisional biopsy has to be avoided are vascular malformations, obviously in order to prevent bleeding (Table I).

In order to expand the surgical field, lateral orbitotomy can be required; in addition, the use of a cryostat can facilitate the procedure, as illustrated in Fig. 2. Lateral

orbitotomy permits us to obtain a wide surgical field, with a minimal number of complications; the bone flap can be removed or dislocated and, after tumor removal, fixed with titanium plates¹². Even if described, we avoid canthotomy using this approach, reducing complications ²². We identified 15 cases of OVM (24.19%): this data is consistent with the literature since vascular malformations represent the most common benign orbital tumor in adults ^{12,14} and usually appears as a well-defined intraconal mass ². All cases were treated without prior embolization by complete surgical removal and transconjunctival and upper eyelid skin crease incisions were the most common surgical approaches.

Idiopathic orbital inflammation (IOI) is a benign inflammatory condition that accounts for approximately 8%-10% of all orbital mass lesions; it has been classified into categories, including anterior, diffuse, posterior or apical, myositis, and dacryoadenitis. Other rare types of IOI include episcleritis, neuritis, and focal masses ²³.

When idiopathic inflammatory pseudotumor is suspected, since it is a diagnosis of exclusion, clinical history has to be carefully investigated: in particular, rheumatic disease, systemic lupus erythematosus (SLE), polyarteritis nodosa, thyroid abnormalities should be noted 14,24. The mainstay of IOI therapy is systemic corticosteroids, which seem to be effective as sole treatment in at least 40% of patients, and in about 60% of patients when combined with other immunosuppressants. The need for orbital biopsy is judicial. Some prefer to obtain a biopsy in many accessible cases, while others reserve biopsies for nonresponding or refractory cases. Surgical debulking or resection can be used in patients that do not respond to systemic corticosteroids, in particular in the case of focal mass IOI, in some anterior/diffuse IOI cases, and in dacryoadenitis ^{25,26}. In our series, 2 patients were affected by chronic dacryoadenitis (16.6% of the IOI group). Dermoid cyst is a common benign congenital tumor, often detected in the first two years of life; it rarely presents in adulthood, most often arising close to the frontozygomatic suture, sometimes with a "dumbbell" configuration, sometimes entirely within the orbit ¹⁴.

Dermoid cysts are classified as superficial (that usually present in younger patients) and deep (typical of adulthood)²⁷. We identified eight patients suffering from deep dermoid cyst (12.9%). Since complete excision is the treatment of choice, surgical removal was performed by upper eyelid skin crease incision in four cases, while the rest of sample underwent a more extended approach, i.e. lateral orbitotomy with coronal scalp flap. In this last case, a more complex approach was performed in order to prevent relapse. In fact, sometimes it is difficult to obtain a complete resection due to bone adherences that can lead to capsule rupture ²⁸. On the other side, enucleation is contraindicated in case of suspected primary malignant tumors or lymphoproliferative disorders: in the first case, it is due to the fact that we have to plan a radical resection or a targeted therapy, while in the second one, it is useful to maintain a residual disease to evaluate response to treatment.

We can distinguish between lymphoma of the orbit (ocular adnexa) and primary ocular lymphoma. Orbital lymphomas present bilaterally in about 10% of cases and seem to be related to chronic ocular infections and thyroid eye disease 28,29. They include marginal zone lymphoma, diffuse lymphoplasmacytic lymphoma, follicular center lymphoma, diffuse large B-cell lymphoma, mantle cell lymphoma, chronic lymphocitic leukemia. The most common signs are exophtalmos, epiphora, soreness, and diplopia ^{28,30}. According to the Surveillance, Epidemiology, and End Results (SEER), there has been a significant increase in the overall incidence of orbital, conjunctival, and lacrimal gland tumors, with lymphoma having the highest incidence³¹. In Italy, Bonavolontà et al. 32, analyzing 2480 orbital space-occupying lesions, found that the most common benign lesions were dermoid cysts (14%) and cavernous hemangiomas (9%). In the same series, non-Hodgkin lymphoma was the most common malignant neoplasm (12%). Lacrimal gland lesions were benign in 64% of cases, whereas the most common malignant tumor was adenoid cystic carcinoma (18%) ³². In our series, we describe 13 patients (21%)with ocular adnexal lymphoma, a malignant lymphoproliferative tumor involving the eyelid, conjunctiva, orbit, and lacrimal gland, almost exclusively extranodal B-cell non-Hodgkin lymphoma². In particular, 6 were cases of extranodal marginal zone lymphoma of mucosaassociated lymphoid tissue (MALT lymphoma), three lymphoplasmacytic lymphoma, two cases of follicular center cell lymphoma, and 1 case of diffuse large B-cell lymphoma. They preferentially affected older patients (in our series, mean age was 71 years), and most of them (69%) presented with exophthalmos and/or epiphora; in all cases, an incisional biopsy was performed, using a "skin-crease" incision in 61.5%, and a transconjunctival approach in the remaining fraction. Incisional biopsy is indicated in case of suspected metastasis too; secondary lesions from solid malignancies into the extra-ocular musculature are very rare and have been reported only sporadically. The frequency of orbital metastases in oncological patient samples varies from 0.07% to 4.7%³³⁻³⁵. Four patients in our study were affected by metastases (6.45%), mean age 58 years +/- 6.06 SD. Females resulted more affected than males (75% vs. 25%), and 75% of patients presented an intraconal lesion. According to the literature, in women breast cancer is the most frequent of malignancies metastasizing into the orbit. One male patient presented a metastasis from kidney cancer. Clinically, all patients presented proptosis, 75% presented motility impairment, 50% pain, and 25% epiphora. Surgical resection is indicated in very selected cases: in fact, it was performed only on a patient affected by clear cell renal carcinoma, using a lateral orbitotomy approach, while 3 patients underwent a surgical biopsy through skin crease incision.

Conclusions

The orbit is an "interdisciplinary" region, being an area of interest for many specialties; orbital lesions may be of different origins, and signs and symptoms are not specific, so that histological diagnosis is required in most cases and can be achieved performing incisional biopsy or complete surgical removal. In every single case, accurate surgical planning is mandatory in order to prevent functional and/or aesthetic complications.

Riassunto

L'obiettivo principale di questo articolo era descrivere la nostra esperienza nella gestione delle lesioni che coinvolgono l'orbita, analizzando segni e sintomi e descrivendo i possibili approcci chirurgici. Le lesioni orbitarie rappresentano un argomento di confine tra splancnocranio e neurocranio.

In questo studio retrospettivo descriviamo la nostra esperienza nella gestione di 62 pazienti con diagnosi di tumori orbitari senza coinvolgimento extraorbitario o intracranico, trattati tra gennaio 2013 e dicembre 2016. Caratteristiche demografiche, sintomi, reperti clinici, tipi istologici, approcci chirurgici sono stati registrati. Nel campione incluso le manifestazioni cliniche più comuni erano esoftalmo e compromissione del movimento oculare; i tipi istologici più frequenti sono risultati essere le malformazioni vascolari ed i linfomi degli annessi oculari. L'asportazione completa della lesione è stata eseguita in 36 pazienti.

La biopsia è stata eseguita in 26 pazienti ed ha confermato la diagnosi di malignità in 16 pazienti (13 linfomi degli annessi e 3 metastasi) e la diagnosi di lesione infiammatoria in 10 pazienti.

In tutti i pazienti l'approccio chirurgico programmato ha permesso di ottenere una diagnosi istologica di certezza e non sono state identificate complicanze/sequele chirurgiche nel periodo di follow-up. Gli approcci chirurgici utilizzati sono stati: l'approccio transcongiuntivale in 19 pazienti (il 30,6% del campione) l'incisione cutanea per la blefaroplastica superiore in 30 pazienti (48,3% del campione) di questi in 9 pazienti l'incisione sovrapalpebrale è stata associata all'orbitotomia laterale ed infine in 13 pazienti è stato utilizzato l'approccio coronale con orbitotomia laterale.

Le neoplasie primarie che coinvolgono l'orbita comprendono un ampio spettro di patologie difficili da gestire senza una diagnosi istologica di certezza.

zVengono descritti diversi accessi chirurgici per eseguire la biopsia incisionale o la resezione della massa che dipendono sia dalla dimensione della lesione, dalla posizione e dal sospetto clinico. In conclusione, quando si approcciano lesioni del contenuto orbitario, è obbligatoria un'accurata pianificazione chirurgica, al fine di prevenire complicanze funzionali e/o estetiche.

References

1. Alsalamah AK, Maktabi AM, Alkatan HM: *Adult orbital lesions in saudi arabia: A multi-centered demographic study with clinico-pathological correlation.* J Epidemiol Glob Health, 2020; 10(4):359-66. doi: 10.2991/jegh.k.200720.001. Epub 2020 Jul 29. PMID: 32959608; PMCID: PMC7758843.

2. Honavar SG, Manjandavida FP: *Recent advances in orbital tumors. A review of publications from 2014-2016.* Asia Pac J Ophthalmol (Phila), 2017; 6(2):153-58. doi: 10.22608/APO. 201736. PMID: 28399335.

3. Mombaerts I, Ramberg I Coupla:nd SE, Heegaard S: *Diagnosis of orbital mass lesions: clinical, radiological, and pathological recommendations.* Surv Ophthalmol. 2019; 64(6):741-56. doi: 10.1016/j.survophthal.2019.06.006. Epub 2019 Jul 2. PMID: 31276737.

4. Terenzi V, Cassoni A, Zadeh OR, Raponi I, Della Monaca M, Bartoli D, Battisti A, Valentini V: *Metastases to oro-maxillo-facial region from distant sites: Are they so rare? A single centre 8-years experience.* Ann Ital Chir, 2015; 86(1):5-8. PMID: 25818917.

4. Papadopoulos GO, Charitonidi E, Filippou N, Fanidou D, Filippou D, Scandalakis P: *Primary basosquamous carcinoma of the lower eyelid with ocular invasion. A case report.* Ann Ital Chir, 2018; 7:S2239253X18028268. PMID: 29626183.

5. Lombardo N, Ferrise P, Piazzetta GL, Bruzzichessi D, Testa D, Viola P, Pelaia C, Motta G: *Maxillary sinus mucocele with orbital complications*. Ann Ital Chir, 2018; 7:S2239253X18028190. PMID: 29661986.

6. Neems L, Echalier EL, Subramanian PS: Orbital tumors and inflammatory disorders: Diagnosis and management. Int Ophthalmol Clin, 2018; Spring 58(2):181-95. doi: 10.1097/IIO.000000 0000000217. PMID: 29517650.

7. Davies BW, Hink EM, Durairaj VD: *Transconjunctival inferi*or orbitotomy: indications, surgical technique, and complications. *Craniomaxillofac trauma reconstr.* 2014; 7(3):169-74. doi: 10.1055/s-0034-1374063. Epub 2014 May 22. PMID: 25136405; PMCID: PMC4130752.

8. Markiewicz MR, Bell RB: *Traditional and contemporary surgical approaches to the orbit.* Oral Maxillofac Surg Clin North Am, 2012; 24(4):573-607. doi: 10.1016/j.coms.2012.08.004. PMID: 23107428.

9. Marenco M, Terenzi V, Pucci R, Lambiase A, Valentini V: *Neuronavigation and intraoperative imaging system in orbital tumor surgery: A review of recent literature.* Ophthalmol Clin Res, 2019; 2(1): 42-47.

10. Dasgupta R, Fishman SJ: *ISSVA classification*. Semin Pediatr Surg, 2014; 23(4):158-61. doi: 10.1053/j.sempedsurg, 2014.06.016. Epub 2014 Jun 19. PMID: 25241091.

11. Colletti G, Biglioli F, Poli T, Dessy M, Cucurullo M, Petrillo M, Tombris S, Waner M, Sesenna E: *Vascular malformations of the orbit (lymphatic, venous, arteriovenous): Diagnosis, management and results.* J Craniomaxillofac Surg, 2019; 47(5):726-40. doi: 10.1016/j.jcms.2018.09.009. Epub 2018 Sep 20. PMID: 30770258.

12. Hayek G, Mercier P, Fournier HD: *Anatomy of the orbit and its surgical approach*. Adv Tech Stand Neurosurg, 2006; 31:35-71. doi: 10.1007/3-211-32234-5_2. PMID: 16768303.

13. Markowski J, Jagosz-Kandziora E, Likus W, Pająk J, Mrukwa-

Kominek E, Paluch J, Dziubdziela W: *Primary orbital tumors: A review of 122 cases during a 23-year period: A histo-clinical study in material from the ENT Department of the Medical University of Silesia.* Med Sci Monit. 2014; 20:988-94. doi: 10.12659/MSM.890433. PMID: 24930391; PMCID: PMC4068967.

14. Chang JR, Gruener AM, McCulley TJ: Orbital disease in neuro-ophthalmology. Neurol Clin, 2017; 35(1):125-44. doi: 10.1016/j.ncl.2016.08.011. PMID: 27886890.

15. Pagni F, Jaconi M, Smith AJ, Brenna A, Valente MG, Leoni S, Leni D, Vacirca F, Sozzi D: *The role of fine needle aspiration of orbital lesions: A case series.* Acta Cytol, 2016; 60(1):31-8. doi: 10.1159/000444385. Epub 2016 Mar 18. PMID: 26986454.

16. Tang W, Hei Y, Xiao L: *Recurrent orbital space-occupying lesions: a clinicopathologic study of 253 cases.* Chin J Cancer Res, 2013; 25(4):423-9. doi: 10.3978/j.issn.1000-9604.2013.08.04. PMID: 23997529; PMCID: PMC3752348.

17. Rose GE, Wright JE: *Pleomorphic adenoma of the lacrimal gland.* Br J Ophthalmol, 1992; 76(7):395-400. doi: 10.1136/bjo.76.7.395. PMID: 1320923; PMCID: PMC504299.

18. Becelli R, Carboni A, Cassoni A, Renzi G, Iannetti G: *Pleomorphic adenoma of the lachrymal gland: Presentation of a clinical case of relapse.* J Craniofac Surg, 2002; 13(1):49-52. doi: 10.1097/00001665-200201000-00010. PMID: 11886993.

19. Tang SX, Lim RP, Al-Dahmash S, Blaydon SM, Cho RI, et al.: *Bilateral lacrimal gland disease: clinical features of 97 cases.* Ophthalmology, 2014; 121(10):2040-6. doi: 10.1016/j.ophtha.2014.04.018. Epub 2014 Jun 3. PMID: 24907059.

20. Chawla B, Kashyap S, Sen S, Bajaj MS, Pushker N, Gupta K, Chandra M, Ghose S: *Clinicopathologic review of epithelial tumors of the lacrimal gland*. Ophthalmic Plast Reconstr Surg, 2013; 29(6):440-5. doi: 10.1097/IOP.0b013e31829f3a0c. PMID: 24145905.

21. Hamed-Azzam S, Verity DH, Rose GE: Lateral canthotomy orbitotomy: A rapid approach to the orbit. Eye (Lond), 2018; 32(2):333-37. doi: 10.1038/eye.2017.173. Epub 2017 Sep 1. PMID: 28862259; PMCID: PMC5811693.

22. Yeşiltaş YS, Gündüz AK: *Idiopathic orbital inflammation: Review of literature and new advances*. Middle East Afr J Ophthalmol, 2018; 25(2):71-80. doi: 10.4103/meajo.MEAJO_44_18. PMID: 30122852; PMCID: PMC6071347.

23. Pakdaman MN, Sepahdari AR, Elkhamary SM: Orbital inflammatory disease: Pictorial review and differential diagnosis. World J Radiol, 2014; 6(4):106-15. doi: 10.4329/wjr.v6.i4.106. PMID: 24778772; PMCID: PMC4000606.

24. Yeşiltaş YS, Gündüz AK: *Idiopathic orbital inflammation: review of literature and new advances.* Middle East Afr J Ophthalmol, 2018; 25(2):71-80. doi: 10.4103/meajo.MEAJO_44_18. PMID: 30122852; PMCID: PMC6071347.

25. Woolf DK, Ahmed M, Plowman PN: *Primary lymphoma of the ocular adnexa (orbital lymphoma) and primary intraocular lymphoma.* Clin Oncol (R Coll Radiol). 2012; 24(5):339-44. doi: 10.1016/j.clon.2012.03.001. Epub 2012 Apr 20. PMID: 22521959.

26. Dave TV, Gupta Rathi S, Kaliki S, Mishra D: Orbital and periorbital dermoid cysts: Comparison of clinical features and management outcomes in children and adults. Eur J Ophthalmol, 2020; 16:1120672120964686. doi: 10.1177/1120672120964686. Epub ahead of print. PMID: 33198489. 27. Knani L, Gatfaoui F, Krifa F, Mahjoub H, Daldoul N, Ben Hadj Hamida F: Les kystes dermoïdes orbitopalpébraux: Étude clinique et résultats thérapeutiques [Orbital dermoid cysts: Clinical spectrum and outcome]. J Fr Ophtalmol, 2015; 38(10):950-4. French. doi: 10.1016/j.jfo.2015.02.012. Epub 2015 Nov 10. PMID: 26563840.

28. Bessell EM, Henk JM, Wright JE, Whitelocke RA: Orbital and conjunctival lymphoma treatment and prognosis. Radiother Oncol, 1988; 13(4):237-44. doi: 10.1016/0167-8140(88)90218-6. PMID: 3217539.

29. Jenkins C, Rose GE, Bunce C, Cree I, Norton A, Plowman PN, Moseley I, Wright JE: *Clinical features associated with survival of patients with lymphoma of the ocular adnexa*. Eye (Lond); 2003; 17(7):809-20. doi: 10.1038/sj.eye.6700379. PMID: 14528242.

30. Ahmed OM, Ma AK, Ahmed TM, Pointdujour-Lim R: *Epidemiology, outcomes, and prognostic factors of orbital lymphoma in the United States.* Orbit, 2020; 39(6):397-402. doi: 10.1080/01676830.2019.1704032. Epub 2020 Jan 2. PMID: 31894706.

31. Bonavolontà G, Strianese D, Grassi P, Comune C, Tranfa F, Uccello G, Iuliano A :*An analysis of 2,480 space-occupying lesions of the orbit from 1976 to 2011*. Ophthalmic Plast Reconstr Surg, 2013; 29(2):79-86. doi: 10.1097/IOP.0b013e31827a7622. PMID: 23470516.

32. Surov A, Behrmann C, Holzhausen HJ, Kösling S: *Lymphomas and metastases of the extra-ocular musculature*. Neuroradiology, 2011; 53(11):909-16. doi: 10.1007/s00234-011-0873-z. Epub 2011 May 3. PMID: 21538045.

33. Magliozzi P, Strianese D, Bonavolontà P, Ferrara M, Ruggiero P, Carandente R, Bonavolontà G, Tranfa F: *Orbital metastases in Italy*. Int J Ophthalmol, 2015; 8(5):1018-23. doi: 10.3980/j.issn. 2222-3959.2015.05.30. PMID: 26558220; PMCID: PMC4630982.

34. Terenzi V, Cassoni A, Zadeh OR, Raponi I, Della Monaca M, Bartoli D, Battisti A, Valentini: *Metastases to oro-maxillo-facial region from distant sites: Are they so rare? A single centre 8-years experience.* Ann Ital Chir, 2015; 86(1):5-8. PMID: 25818917.