MANAGEMENT OF ORBITAL TUMORS PRESENTING AS UNILATERAL EXOPHTHALMOS

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ABSTRACT

Aim of the study. We aim to present our experience regarding the management of exophthalmos as the first clinical manifestation of a space-occupying orbital lesion. Material and methods. We performed a retrospective review, selecting 17 patients with orbital tumors that initially presented for proptosis. Results. Following multidisciplinary clinical and imaging evaluation, three primary orbital tumors were diagnosed, one mass pertaining to the lacrimal gland, and 13 secondary tumors. Surgical treatment was performed in 11 patients. Complete excision was performed in three benign tumors, complete removal and orbital content preservation was achieved in eight patients, orbital exenteration was necessary in two cases of orbital malignancies, and extended orbital exenteration was performed in one patient. Four cases did not have indication of surgery due to internal carotid artery invasion or extensive intracranial invasion. Two patients were diagnosed with lymphoma following biopsy and underwent oncologic treatment. Conclusion. An early diagnosis of an orbital mass presenting as proptosis involves multidisciplinary evaluation for the exclusion of other causes of exophthalmos. Orbital content and function preservation are the main goals of the surgical treatment.

Key words: orbit, tumor, exophthalmos, proptosis, exenteration

INTRODUCTION

Tumors growing inside the orbital space are extremely diverse considering the complex anatomy and the multitude of neighboring structures as possible origin for tumor development. The particularity of orbital lesions is related to their location at the crossroads of multiple specialties: Maxillofacial surgery, ENT, Ophthalmology, Neurosurgery. As a consequence, the first presentation of the patient will be dependent on the initial signs and symptoms and the diagnosis as well as treatment will be multidisciplinary. Proptosis can be the initial manifestation of the disease in up to half of the orbital tumor patients. For an accurate diagnosis and treatment plan, other causes of exophthalmos must be excluded, such as inflammatory, traumatic or thyroid disease [1, 2]. The majority of space-occupying lesions of the orbit are benign, with malignant tumors accounting for only one third of orbital tumors. The most common benign primary tumors of the orbit are cavernous hemangiomas and dermoid cysts, while the most prevalent primary malignancy is represented by lymphoma. Extended tumors to the orbit are most often basal cell and squamous cell carcinomas. Orbital metastasis most often comes from breast carcinoma, followed by unknown primary tumor, kidney and lung carcinoma [3, 4, 5]. Almost half of all orbital tumors are encountered in the extracranial area, with the rest found intraconally and in the area of the lacrimal gland [6, 7].

MATERIAL AND METHODS

We performed a retrospective review from 2012 to 2016, selecting a total of 17 patients with orbital tumors, nine men and eight women, aged between 23 and 74. Only patients that first presented for exophthalmos were included. All aspects related to diagnosis and surgical treatment were evaluated.
RESULTS

Proptosis was the presenting sign in all cases. In nine patients proptosis was the only complaint. 16 patients presented with unilateral exophthalmos and one patient with bilateral exophthalmos. Additional complaints were: diplopia (3), nasal obstruction (6), epistaxis (3), cephalalgia (5), visual disturbances (3), and hemi-facial algia (4).

The initial investigations included a multidisciplinary approach involving ophthalmology consultation in all cases, neurosurgical examination in 13 cases and ENT evaluation in seven cases.

Imaging included orbital ultrasound (3), CT scan (13), IRM (14). There were three primary orbital tumors and one mass pertaining to the lacrimal gland. Secondary tumors extending to the orbit involved five sino-nasal tumors extended to the orbit, six skin tumors with orbital involvement, two ventral skull base tumors with orbital invasion.

Prior to the surgery, biopsy was performed in seven cases, resulting in two squamous cell carcinomas, three basal cell carcinomas and two lymphomas.

Surgical treatment was performed in 11 patients. Four tumors were considered unresectable due to important intracranial invasion (1), or internal carotid artery invasion of the intracranial segment (3). In the two patients with lymphoma the treatment was not surgical.

Tumor removal was performed by complete excision in all benign tumors (3), complete removal and orbital content preservation in eight patients, orbital exenteration was necessary in two cases of orbital malignancies, extended orbital exenteration was performed in one patient. Plasty of the resulting defect was necessary in seven cases by using skin grafts (3), temporalis muscle flaps (2), temporo-parietal flaps (2).

Postoperative histology revealed one mucocele, two hemangiomas, eight squamous cell carcinomas, three basal cell carcinomas, and one adenoid cystic carcinoma.

The postoperative evolution was favourable in all patients with benign lesions, with the resolution of exophthalmos and related symptoms, without evidence of local recurrence. Patients with squamous cell carcinomas and adenoid cystic carcinoma underwent further multimodal treatment, including radiotherapy, chemotherapy and further oncologic surveillance. In patients with basal cell carcinomas the only treatment was excisional surgery. Two of the patients presented with local recurrence at 1 and respectively 1.5 years after the initial surgery. Therefore a second excisional procedure was performed in both cases. Patients diagnosed with lymphomas underwent oncologic treatment and surveillance.

DISCUSSIONS

Space-occupying lesions of the orbit may initially manifest as a variety of signs and symptoms that are more or less common in the medical practice and can therefore cause different degrees of concern [8]. Initial patient referral is important since it could accelerate or delay the diagnosis of an orbital mass considering the frequency of the presentation sign in every-day practice.

Patients in our study initially presented to the ophthalmologist for exophthalmos and associated visual disturbances including diplopia. Other authors revealed different, more common reasons for initial ophthalmologic presentation: the investigation of visual disturbances and the need to prescribe or renew glasses, as well as pain felt in the orbital region [9]. As a presenting sign, proptosis can be the initial manifestation of a wide range of disorders including inflammatory, traumatic, endocrine, tumoral [8, 10]. Therefore, a thorough differential diagnosis was performed in all cases included in the study, in order to exclude non-tumor disorders. This was achieved by the help of a detailed anamnesis, careful clinical examination, proper imaging and interdisciplinary evaluation for establishing the suitability of surgery and
additional treatment. History included detailed information about the onset of proptosis and associated symptoms, especially the presence of inflammatory signs. The clinical examination determined the amount and direction of globe displacement, the ocular movements and the visual function, focusing on the search for the “Six P’s” formula described by May and colleagues: proptosis, pain, progression, pulsation, palpation, and periorbital changes [11].

The assessment of thyroid function was part of the initial investigations performed in our study in all patients with exophthalmos that did not present with obvious external signs suggesting tumor etiology. This investigation was necessary considering the frequency of Basedow-related exophthalmos. Although bilateral exophthalmos is the usual encounter in thyroid gland disease, unilateral exophthalmos is also a possible presentation in the early stages of the disease [12, 13]. Therefore, this differential diagnosis should always be considered and excluded. In our case series we encountered a patient presenting with bilateral proptosis due to an extended tumor located centrally in between the two orbits. Underdiagnoses of an orbital mass in such a case is possible by mistaking the initial presentation as a cause of thyroid gland disease. Thyroid hormone level is a necessary investigation in the assessment of exophthalmos [12, 13, 14] in the absence of an obvious causing factor, such as an extended skin malignancy invading the orbit, when an extensive differential diagnosis is not necessary.

In addition to a thorough clinical examination involving multiple specialties, patients in our study were further investigated by the help of CT and/or MRI. Although CT is mostly recommended due to availability and speed and also the precision in determining bone invasion, some authors prefer MRI for investigating orbital tumors due to the good soft tissue resolution and possibility to better evaluate optic nerve invasion and intracranial extension [9]. Doppler ultrasound is useful in differentiating vascular tumors [15]. In most cases, the corroboration of clinical and imagistic information allows an adequate differentiation between the various types of orbital lesions, especially differentiating between benign and malignant lesions, necessary for further decision of the treatment plan.

The benign lesions encountered in our case series included one mucocele and two hemangiomas that were all treated by complete surgical removal. Although benign tumors involving the orbit have good overall prognosis, the functional and aesthetic consequences of tumor growth and consequent tumor removal can be distressing for the patient. As one of the most common benign lesions involving the orbit, mucoceles can grow to impressive dimensions, leading to bone resorption [16]. This is particularly important when the floor of the orbit is involved. Performing plasty of the defect and reconstruction of the orbital floor using a titanium mesh, or composite regional flaps can help reduce the functional disturbances related to enophthalmos and diplopia [17, 18].

In our study, the indication for surgical treatment was decided following a multidisciplinary evaluation and a thorough clinical and imagistic assessment. A biopsy was performed prior to surgery in cases where the tumor was directly accessible and when there was uncertainty of either malignant or benign character of the growing mass. Some authors underline the importance of intraoperative frozen-section in establishing the extent of surgery and when intraoperative findings suggest a need for a change in the pre-established treatment method [19, 11]. An orbital lesion suggestive for lymphoma should be biopsied preoperatively, since surgical removal is not indicated in such cases [20].

The choice of surgical approach and reconstruction technique must take into account the incidence of postoperative sequelae. Orbital surgery implies an increased risk of injury to important anatomical structures - the extraocular muscles, the optic nerve and other cranial nerves, resulting in functional disturbances. Any change in the volume of the orbital content, due to either
postoperative fibrous tissue formation or surgically removed tissue, will result in permanent diplopia. Postoperative radiotherapy following malignant tumor removal can also accentuate these changes. Optic nerve injury inside the orbit or at the level of the skull base can result in partial or complete vision loss. A postoperative compressive retrobulbar hematoma can also cause damage to the optic nerve due to prolonged ischemia [9]. Functional sequelae related to orbital surgery must be minimized particularly when surgery for benign tumors is performed.

In the choice of surgical approach, we considered that incisions involving the upper or lower eyelid can determine the appearance of postoperative ectropion and lagophthalmos. In the case of such an occurrence, a minimally invasive solution for the correction of ectropion is the use of fat transfer. By this method, the volume of the superior genian region is restored, rendering increased support for the eyelid [21] while also addressing the scar tissue and increasing the pliability of the soft tissues [22]. The same increased eyelid support, as well as restoration of facial symmetry, can be achieved by the insertion of barbed threads for soft tissue suspension at the level of the cheek region [23].

The extent of the surgical procedure is the most important decision considering the functional postoperative sequelae. There is an ongoing debate on the indications of orbital exenteration [24]. The periorbita is viewed by most authors as the anatomical reference landmark in establishing orbital involvement. In our study, the invasion of the periorbital fat, extraocular muscles and orbital apex indicated the need for performing orbital exenteration. Periorbital involvement was investigated by the help of CT or MRI, although intraoperative assessment did not always match the preoperative result of the imaging studies. Some authors recommend a combined CT and MRI assessment for establishing the degree of invasion [1]. We consider that the intraoperative aspect and frozen section results are the most reliable methods of determining periorbital involvement and deciding the extent of surgery. The patient should always be informed preoperatively about the possibility of complete orbital content removal even in cases where conservation is planned [24].

The choice of surgical treatment and defect reconstruction method is most difficult in the case of secondary tumors extending to the orbit. It is stated that malignant tumors of the sinonasal cavity and skull base involve the orbit in 50% to 80% of cases [2]. The incidence of orbital invasion is highest in ethmoid malignancies [25, 26]. The absence of orbital signs and symptoms at presentation is not equivalent with absence of orbital involvement. Imaging studies involving CT and MRI, as well as ophthalmologic examination, are of outmost importance in all cases of malignancies arising from the sinuses and nasal cavity. For adequate access, sinonasal malignancies extending to the orbit encountered in our case series were resected via a modified Weber- Fergusson approach. Other authors describe various transfacial and craniofacial approaches for accurate exposure and resection of extended sino-nasal malignancies [27].

Following surgery for malignant tumors of the maxillary sinus extended to the orbit, the restoration of the postoperative defect requires the manufacturing of an obturator prosthesis for restoring the contours of the midface [28] and providing support for the orbital content until further reconstructive methods by regional or distant flaps are decided. When orbital exenteration is also performed, an episthesis can be used for improving appearance [29]. The plasty of the anophthalmic socket can be performed by the help of a temporalis muscle flap and skin graft. A temporo-parietal fascio-cutaneous flap is also an option for reconstructing the orbital and upper genian region. Local flaps, like the genian flap can also be used for additional adequate closure of the soft tissues. For more extended defects, a free flap might be necessary for bone and soft tissue coverage [30].

A multidisciplinary surgical team, involving a maxillofacial surgeon, a neurosurgeon and an ophthalmologist, was
necessary for the removal of skull base tumors extended to the orbit. Optimal access to the tumor was achieved by a craniofacial approach. In cases of internal carotid artery or cavernous sinus invasion, surgery was not indicated. Another criteria that needed to be considered in the preoperative planning was the presence of extensive intracranial invasion that would increase morbidity and would impede obtaining free oncologic margins. This is consistent with other studies on the subject [2, 30].

CONCLUSIONS
Time is precious in the diagnosis of an orbital tumor and can mean the difference between orbital content preservation and orbital exenteration. Therefore, the management of proptosis as an initial complaint should include multiple specialties and imaging methods for early diagnosis of an orbital mass and consequent surgical treatment aiming at complete tumor removal and function preservation whenever possible.
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