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Case Report
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Recurrent Orbital Myxofibrosarcoma: Case Report and Review of Literature

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Abstract

Myxofibrosarcoma in orbital location is extremely rare. There are less than 10 reported cases of orbital myxofibrosarcoma. The authors report a case of a patient with a previous resected low-grade myxofibrosarcoma in the right orbit, who 16-years later presented with the same tumor. This first recurrence was resected, leaving a residual mass. Two years later, the patient presented with an extensive mass in the right orbit, sinuses, and nasal cavities. The histopathological examination showed high-grade myxofibrosarcoma. The patient underwent palliative radiotherapy and palliative care. The risk of metastasis of this type of tumor depends on the degree of histological differentiation. But local recurrence rate is similar for all with an increase of metastasis risk with every recurrence. Due to the few cases reported there are no standardized management guidelines. However, it is recommended wide surgical margins be associated with postoperative radiotherapy in the presence of residual lesions or high-grade tumors.

Keywords: Myxofibrosarcoma; Orbital; Proptosis

Introduction

Myxofibrosarcoma (MFS) is the most common type of fibroblastic sarcoma in elderly patients. It is a slow-growing mesenchymal tumor that usually occurs in the extremities [1]. Its orbital location is extremely rare as there are few cases described in the literature at the moment of this review. Therefore, there are no treatments guidelines available for this disease, thus the authors present this case of a recurrent orbital myxofibrosarcoma in order to collaborate towards future studies and this orbital tumor management.

Case Presentation

A 73-year-old woman presented with a 5-month painful right proptosis and a same-side exodeviation. Her past medical history was significant for a right orbital mass resection reported by an external hospital as a low-grade myxofibrosarcoma 16 years earlier. Her other medical background included arterial hypertension, dyslipidemia, fibromyalgia, essential tremor, suspended tobacco use, and she had been treated for a depressive episode. The patient was taking atorvastatin and sertraline, at the moment of presentation.

On the first physical examination, she presented with an obvious right exophthalmos and exotropia (Figure 1). Uncorrected visual acuity was 20/150 in the Right Eye (RE) and 20/20 in the Left Eye (LE). Intraocular pressure was normal in both eyes. Slit-lamp examination was unremarkable. Fundus examination revealed a congested right optic disc.

Computerized Tomography scans (CTs) revealed a thrombosed right orbital vascular malformation and Magnetic Resonance Angiography (MRA) revealed an orbital mass with a vascularized component in the internal portion of the right orbit (Figure 2).

Considering the patient's background, a relapse of the original tumor was suspected. Thus, a transcaruncular orbitotomy with excisional biopsy of the lesion was performed. The mass was lobular and solid in appearance, of medial location, and deep extension, almost reaching the orbital apex. Due to the lesion size, it was necessary to widen the surgical field with a vertical division of the upper eyelid at 5 mm from the superior punctum. The resected mass was 50 mm × 30 mm × 19 mm (Figure 3).

Histologic examination revealed fused cells, of elongated, oval, round nuclei, some pleomorphic and hyperchromatic, eosinophilic cytoplasm, mitotic count of 1 to 3 in 10 fields of higher magnification. Stroma with mild to moderate inflammatory elements of lymphocyte type, congestive

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Figure 1: Patient at first consult presenting right exophthalmos and exotropia.

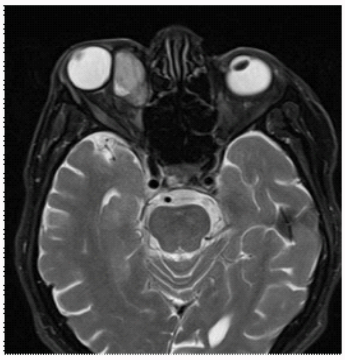

Figure 2: Brain resonance showing a thrombosed right orbital vascular malformation.

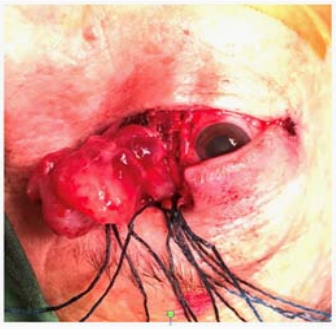

Figure 3: Resected mass.

vascularization, hyalinized fibrosis, myxoid areas. No necrosis was found. Immunohistochemical analysis of the tumor cells was positive for vimentin, desmin, ki-67 positive in 10% of the nucleuses and CD34 in 80% of the cells negative for S100 and actine. These features were suggestive of low-grade myxofibrosarcoma.

Due to the presence of tumor in the surgical margin of the biopsy a post-surgical CT was done, which confirmed the presence of a residual mass. This case was presented to the oncology committee to evaluate the need for exenteration of coadjuvant therapy. Consensus was not reached. Therefore, no further procedures were done.

The patient stayed asymptomatic during medical follow-up for two years, after that period, the patient presented with two-week paresthesia of the right orbital region, progressive exophthalmos, and a recurrent nosebleed from the same side. On examination, the patient showed greater proptosis in the right eye. The evaluation by otolaryngology revealed a nasal cavity mass. Resonance revealed an extensive mass centered in the perinasal cavities of about 60 mm × 50 mm × 62 mm. The tumor occupied the ethmoid cells, maxillary

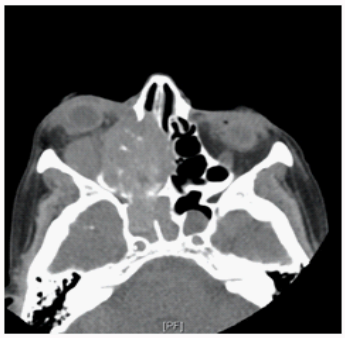

Figure 4: Resonance showing recurrence of the tumor centered in the perinasal cavities.

sinus, and part of the right orbit, with intraconal involvement, and which posteriorly extended to the sphenoid sinus, compressing the cavernous sinus (Figure 4).

Through the nasal passage, a new biopsy was performed. It was diagnosed as high-grade myxofibrosarcoma. The case was presented again to the oncology committee, along the lesion was out of surgical reach. It was decided that palliative care management and palliative radiotherapy was the best option for this patient.

Discussion

Myxofibrosarcoma (MFS), previously called malignant myxoid fibrous histiocytoma, is one of the most common fibroblastic sarcomas in elderly patients (20%). It is a slow-growing mesenchymal tumor that usually occurs in the extremities, and its orbital location is extremely rare [1]. In the literature there are less than 10 reported cases of orbital myxofibrosarcoma, however, it is likely that there is an underreporting due to the change in its nomenclature in recent years [2-10]. In 2002, the World Health Organization modified the classification of MFS, so that part of the previously reported orbital Malignant Fibrous Histiocytomas (MFH) could correspond to the same entity, unlike other subtypes of FMH (pleomorphic, giant cell and inflammatory) that did not represent a defined entity [11,12]. Characteristics of the previous studies are summarized in Supplementary Table 1; however it was not possible to find the full article of the work by Imai et al. [7].

The clinical behavior of myxofibrosarcomas is related to the degree of histological differentiation. Low-grade tumors have a low probability of metastasizing (20%), however the local recurrence rate is similar for all tumor grades, with rates ranging between 17% and 60%, and with each new recurrence the risk of metastasis recurrence increases [1,8,13,14]. The high rates of local recurrence could be explained by the frequent vascular extension of the tumor [15]. Recent studies have shown that the only significant predictor of local recurrence risk is the surgical margin [14]. Risk factors associated with disease-specific mortality are size greater than 5 cm, tumor necrosis, and <75% myxoid component on histology [13]. Compared to other soft tissue sarcomas, MFS have better survival, since a 5-year survival rate is estimated at 64% to 77% in high-grade MFS [16].

Managing these patients is a real challenge. The described infiltrative character makes it very difficult to define the tumor margins, increasing the probability of residual lesions and therefore of local recurrence. In addition, each recurrence increases the probability of having higher-grade lesions, and therefore of metastasizing. The few cases reported to date do not allow for standardized management

guidelines, but in general, resection with wide surgical margins is recommended, associated with postoperative radiotherapy in the presence of residual lesions or high-grade tumors.

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