



Case report

A rare erosive orbital mass in a child: Case report of myofibroma

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Abstract

Purpose: To present the clinical, histological, and radiographic findings of a case of orbital myofibroma in an unusual location. The literature is reviewed and the clinical relevance discussed.

Methods: A 5-year-old boy was examined with a 1.5-month history of progressive swelling in the left supraorbital region.

Results: Examination revealed a firm, painless mass in the supralateral region of the left orbit with slight reddish discoloration of the overlying skin. Computerized tomography (CT) scan images showed a well demarcated, homogenous, solid mass with extension to the lacrimal gland region and adjacent to frontal bone erosion. The mass was surgically excised and was confirmed to be myofibroma in diagnostic histological studies. There has been no evidence of recurrence in the first year after surgery.

Conclusions: Clinical appearance and imaging findings are unspecific for this tumor, and histological examination still remains the definite method of diagnosis. Therefore, it is important to be able to differentiate myofibromas from other malignant tumors with a similar presentation in pediatric patients to avoid mismanagement.

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Keywords: Orbital tumor; Lacrimal gland; Children; Histopathology

Introduction

Although rare, myofibroma is the most common benign fibrous tumor of infancy. There are three different types of myofibroma: solitary, multicentric, and multicentric with visceral involvement. Myofibromas predominantly involve the skin and superficial soft tissue of the head and neck in children especially those younger than two year old.¹ While it rarely involves the ocular region, myofibroma primarily occurs in the extra orbital region and can be with or without bone invasion.^{2,3} Imaging findings on this tumor lack specificity and the

diagnosis has been mainly based on histologic and histochemical inputs.^{1,2,4} Excellent prognosis has been reported for solitary and multicentric without visceral involvement types. Visceral involvement has resulted in poor prognosis (74% mortality rate).¹ Complete surgical excision has mainly been the treatment of choice with a chance of no recurrence.^{1,3} The distinction between benign myofibroma and malignant tumors in pediatric age group appears to be crucial in avoiding mismanagement.

The first objective of this study is to report a rare case of solitary orbital myofibroma with extension to lacrimal gland region and bone erosion. The second objective is to discuss relevant challenges in diagnosis and management.

Case report

A 5-year-old, otherwise healthy male was examined with a history (about one month and a half) of progressive, painless swelling in the left supraorbital region. Family history was

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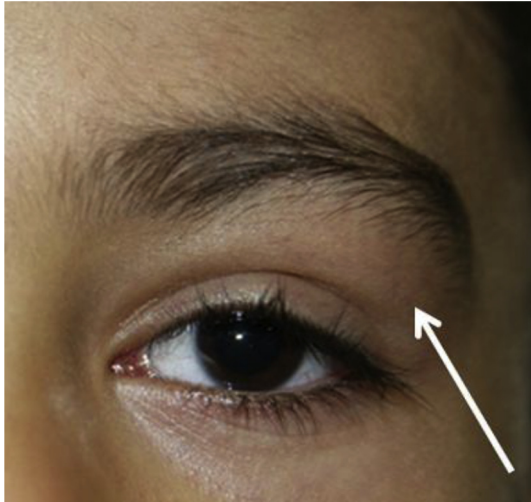


Fig. 1. External examination: 5 years old male with a left upper eyelid mass, and a subtle reddish brown discoloration of the overlying skin (Arrow).

negative for genetic disorders, tumors, or ocular problems. There was no history of trauma. No abnormality was found on general examination. The patient had normal visual acuity and pupillary function in both eyes without proptosis or limitations in eye movements.

The mass was firm, painless, and located in the left upper eyelid with a reddish brown discoloration of the overlying skin (Fig. 1). The examination of globe was normal in both eyes.

Computerized tomography (CT) scan images demonstrated a well-circumscribed (approximately 2×2 cm), homogenous, and isodense tumor with irregular borders in the supra-temporal part of the left orbit with extension to lacrimal gland site. While extensive frontal bone erosion (supralateral) was observed, no globe compression or muscle involvement was detected (Fig. 2).

The following differential diagnoses were considered for this particular case: a) benign tumors e.g. Langerhans cell histiocytosis, lymphangioma, glioma, and plexiform neurofibroma, and b) malignant tumors e.g. neuroblastoma, rhabdomyosarcoma, and leukemic masses.

The mass was fully excised through anterior orbitotomy. It did not involve lacrimal gland and had no extension to the cranium or sinuses.

Macroscopic examination revealed two pieces of creamy tissue with a rubbery consistency. Microscopic examination showed a neoplastic tissue composed of bland looking spindle cell proliferation with a hemangiopericytoma like vascular pattern. An intervening mature bone tissue was also observed. Mitotic figures were scant, and no sign of necrosis was observed. The immunohistochemistry (IHC) study was positive for smooth muscle actin (SMA) and revealed negative results for Bcl2, CD34 (only positive in blood vessels) and alkaline phosphatase (ALK). Ki67 was noted in less than 1-2% of the cells (Fig. 3).

Benign myofibroma was diagnosed based on histopathology and immunohistochemical studies. Systematic imaging followed the diagnosis without evidence of visceral involvement.

The early postoperative period was uneventful. One year follow-up examination indicated no recurrence on imaging.

Discussion

Myofibromas are generally a group of rare, benign, fibromatous tumors that mostly affect children. The appearance can sometimes mimic more aggressive and malignant tumors. In those scenarios the scarcity of myofibroma creates a diagnostic challenge that may lead to a misdiagnosis and subsequent inappropriate patient management. The diagnosis is

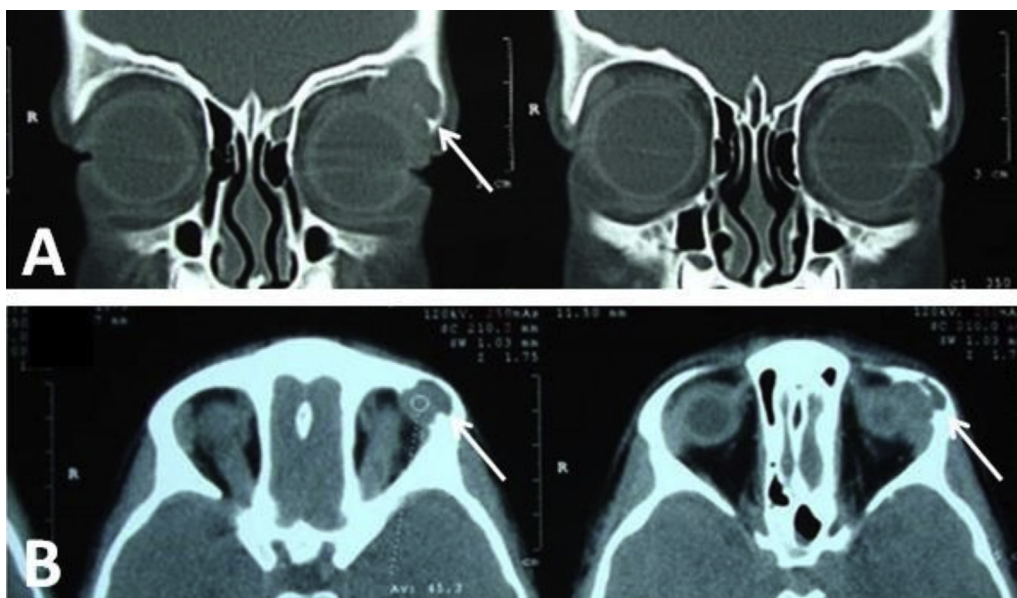


Fig. 2. Orbital computed tomography: A. Coronal view demonstrated an isodense homogeneous well circumscribed mass with irregular borders in the lateral and superior parts of the left orbit with extension to lacrimal gland site with extensive supralateral frontal bone erosion (Arrow). B. Axial view of the findings described (Arrow).

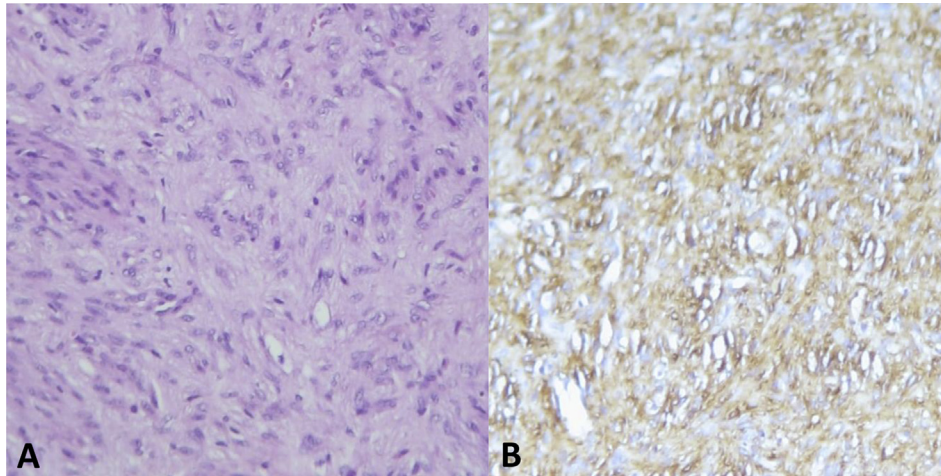


Fig. 3. Histologic features and immunostaining findings of the tumor: A) Interweaving fascicles of myoid-appearing spindle cells with elongated nuclei and abundant eosinophilic cytoplasm (H&E, $\times 100$). B) The cells show diffuse and strong positivity for smooth muscle actin (SMA) (Immunostaining, $\times 100$).

predominantly made on the basis of histological studies and immunohistochemical staining.^{1–5}

This case report and conducted literature review focus on the clinical presentation, histologic features, immunohistochemical profile, and therapeutic management of a solitary orbital myofibroma. To the best of our knowledge and the extent of our literature review, the present case is among rare reported cases located at the superolateral orbit.^{1,2,4} Additionally, the involvement of both bone and soft tissue was a unique characteristic of the current case. In Iran, there has been only one other report of myofibroma in the orbital region that was in the eyelid.⁶

The tumor was first described by Stout in 1954.⁷ Since then and until 1981, it had been referred to with various names. Later, it was renamed to myofibroma/myofibromatosis by Chung and Enzinger.⁸ Orbital involvement represents a rare presentation of this tumor. Kodsí et al, reported only one case of myofibroma in a review of 340 orbital tumors in children, over a period of sixty years.⁹

Myofibroma mainly develops in children younger than 2 year old (89%), and in many cases it was present at birth (54%).¹ It mostly occurs in the head and neck region and the presentation can be as a solitary (74%) or multifocal (26%) tumor.⁵ The multifocal form involves skin, subcutaneous tissue, skeletal muscle, and bone, and generally has a favorable prognosis (multicentric). It can also have generalized visceral involvement with characteristically poor prognosis.^{4,10} Myofibroma mostly occur sporadically, however, autosomal recessive and dominant presentations have also been suggested.¹

The solitary orbital myofibroma has a higher prevalence among male patients and develops in younger ages. There is a tendency for this tumor to involve the left eye; however, no explanation has been presented yet.¹ It is also shown that orbital myofibromas are more frequently located in the lower orbital wall and predominantly involve orbital bones, compared with orbital soft tissue.¹ Patients mostly present with a mass lesion involving the orbit or eyelid, likely accompanied by a sensation of retrobulbar pressure as well as

proptosis or limitation in eye movements. On examination, tumors are usually described as firm, painless, and well-circumscribed lesions.¹

Findings from CT scans are not specific and are often unreliable for definite diagnosis. Myofibromas usually appear as heterogeneous and well-circumscribed masses with moderate vascularity.^{3–5} The presence of ominous signs like bone erosion can notably make the diagnosis more difficult. There is also a rare chance for the tumor to extend to adjacent orbital structures, like extraocular muscles,⁷ lacrimal gland and sinuses, or even the cranium.¹¹

Histological findings are specific for diagnosis. Myofibromas appear as biphasic tumors, showing whorled periphery and nodular areas of fusiform cells. Extracellular collagen, mixed with a central population of small, primitive appearing and darkly staining cells, has a rich vascular network yielding an appearance similar to hemangiopericytoma. Central necrosis and mitoses are occasionally seen. A small subset of tumors may display atypical features which, according to Linos K et al, do not appear to adversely affect the prognosis.¹² Myofibromas are strongly positive for SMA and vimentin in IHC studies, but are usually negative for muscle specific actin, desmin, and CD34.^{1,12,13}

Overall, these lesions have excellent long-term clinical prognosis. Although there is a possibility for lesions to undergo spontaneous involution or regression with time, surgical excision still remains as the treatment of choice. Mynatt et al, on a review of 24 cases of myofibroma with a mean follow-up of 34.6 months, reported two recurrences at months 4 and 6 in patients following surgical excision.¹ Persaud reported no recurrences in nine patients that had undergone surgical excision (7 complete excisions with a follow-up period ranging from 4 to 6 months and 2 partial excisions with 7 years of follow-up).³

From clinical, imaging, and histopathological perspectives, the current case was similar to other reported cases.^{1–14} It appeared to be among rare cases in terms of location in the superolateral orbital area^{1–5} and had similar extents of soft tissue and bone involvement. Other cases predominantly

involved either bone (interosseous) or soft tissue (no bone erosion). The case presented by Larsen et al,² predominantly involved soft tissue (no adjacent bone erosion) and cases presented by Mynatt et al,¹ and Rodrigues et al,⁴ were mainly interosseous tumors. Campbell et al also reported a case of juvenile fibromatosis with a similar presentation, in terms of location and imaging. However, the report did not provide IHC data on the specimens.¹⁴

The postoperative follow-ups did not show any sign of tumor recurrence, well-aligned with prior relevant reports. The latter is anticipated to be largely due to the complete excision of the tumor and the solitary aspect of it.^{1,3}

In conclusion, the lesion can resemble a wide range of benign and malignant tumors especially in older children. Considering that a complete excision can cure the tumor with a very low rate of recurrence, definite diagnosis and differentiation from other tumors by microscopic findings and immunohistochemical staining is mandatory.

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