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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 scan images showed a well demarcated, homogenous, solid mass with extension to the lacrimal gland region and adjacent 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.

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 years 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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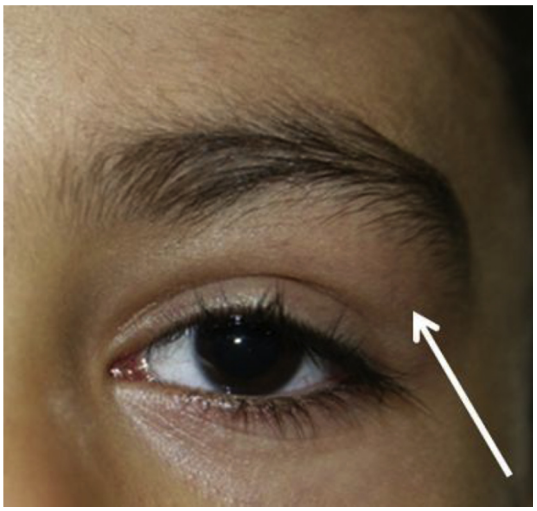

Fig. 1. External examination: 5 years old male with a left upper eyelid mass, and a subtle reddish brown discoloration of the overlying skin (Arrow).

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 boarders 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 indentation 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 than1-2% 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

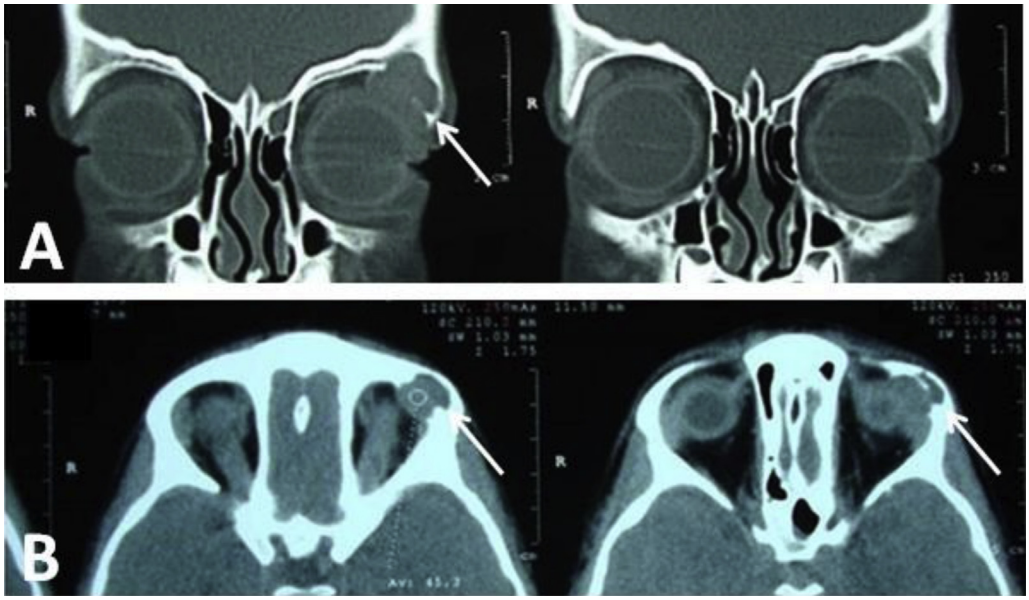

Fig. 2. Orbital Computed tomography: A. Coronal view demonstrated an isodense homogeneous well circumscribed mass with irregular boarders in the lateral and superior parts of the left orbit with extension to lacrimal gland site with extensive supralateral frontal bone erosion (Arrow). B. Axial view of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

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