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Intraosseous Myofibroma of the Zygomatic Bone in Pediatric Patient: A Case Report

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Abstract: *Myofibroma and myofibromatosis are terms used to denote the solitary (myofibroma) or multicentric (myofibromatosis) occurrence of benign neoplasms composed of contractile myoid cells arranged around thin-walled blood vessels. A case is reported of a 7-year-old female patient who presented with a solitary intraosseous myofibroma in the inferolateral aspect of the left orbit. This article describes the clinical, radiographic and histopathological features of an intraosseous myofibroma in a female child. Solitary myofibromas are rare in the orbit. Their rapid growth and bony destruction can mimic malignant tumors. Complete excision with close follow-up is the preferred treatment. Solitary myofibroma should be considered in the differential diagnoses of fibrous tumors with bone destruction in the orbit.*

Keywords:*Intraosseous, zygoma, Myofibroma* **.**

I. 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]

Imaging findings on this tumor lack specificity and the diagnosis has been mainly based on histologic and his to chemical 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]

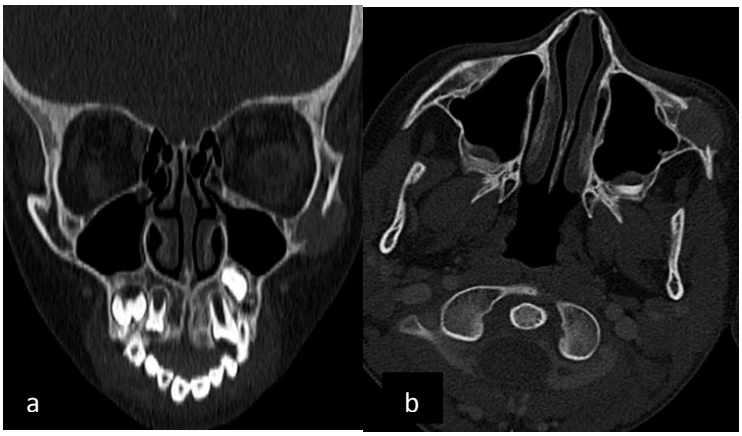
The objective of this study is to report a rare case of solitary orbital myofibroma that confirmed by his to pathological study.

II. CASE REPORT

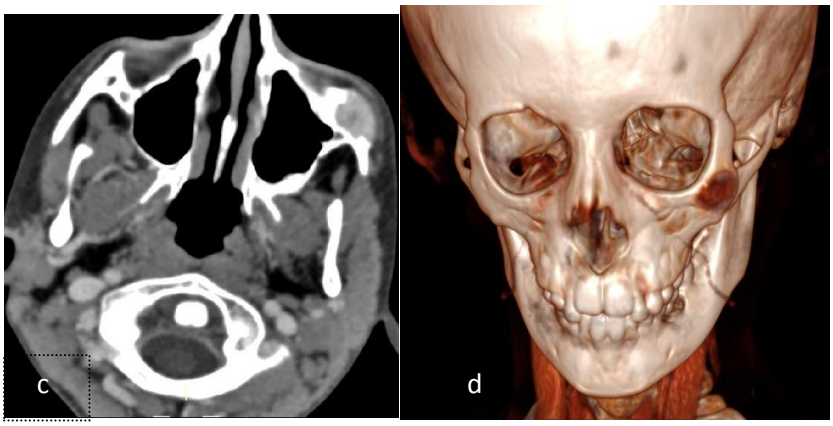
A 7-year-old female presented with a swelling on left side of the face mainly over the cheek area that had enlarged progressively during the preceding 4 months. There was no history of discharge, pain, fever, numbness or paraesthesia. Family history was negative for genetic disorders, tumors, or ocular 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. The patient was moderately built and well-nourished, and no gross physical or mental abnormalities were detected on doing thorough general physical examination.

Computerized tomography (CT)scan images demonstrated small (approximately2 × 1.9 cm), lytic, expansile bony lesion involving the left zygomatic bone, situated at the inferolateral aspect of left orbit (fig 1). It associated with heterogenous enhancing soft tissue component after contrast study (fig 2). There was no intra-orbital extension of the lesion. Intra-orbital structures and rest of the bony orbital walls appeared normal.



(Figure1):Axial (a)and coronal(b) imagesof the orbit shows a well-defined lytic lesion with evidence of bony erosion at the inferolateral aspect of left orbit.



(Figure 2):Axial images (c)with contrast shows heterogenous enhancing soft tissue component.

3D images(d) demonstrate the exact location of the zygomatic lesion.

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.

But in this case the radiological diagnosis of Langerhans cell histiocytosis was suspected but after complimentary radiological studies, no other bony lesions could be identified.

The lesion was surgically explored. After blunt dissection in infraorbital region, an area of surface bony erosion was exposed. On palpation, it was a soft tissue lesion attached to the surrounding bony cavity. Excision was done, and the specimen was sent for histopathological examination.

Histopathology of the excised tissue showed that the tumor was composed of spindle cells that arranged in fascicles and whorls. Stroma was fibrous and myxomatous with basophilic appearance. Focal increased vascularity with hemangiopericytoma like the pattern was seen. There was no evidence of tuberculosis and malignancy. An impression of intraosseous myofibroma was given

III. DISCUSSION

The tumor was first described by Stout in 1954. [6] Since then and until 1981, it had been referred to with various names. Later, it was renamed to myofibroma/myofibromatosis by Chung and Enzinger. [7] Orbital involvement represents a rare presentation of this tumor. Kodsi et al, reported only one case of myofibroma in a review of 340 orbital tumors in children, over a period of sixty years [.8]

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 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]

Our case was a case of solitary myofibroma involving the calvarial bone in a female child although literature reveals a slight male predilection

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, 4, 5]

Radiographically, intraosseous myofibromas are well-circumscribed, osteolytic in nature with sclerotic margins. If a calvarial lesion is present, computed tomography scanning may reveal a lytic lesion causing expansion of the inner and outer tables. Central areas of calcification may also be present. No such calcifications were evident in the present case.

The diagnosis is usually established by excisional biopsy. The use of fine-needle aspiration biopsy has been described but is not well established. Microscopically, the lesions are characterized by fascicles, whorls, and nodules of spindle-shaped cells with myofibroblastic features. [9].

Treatment of myofibroma is usually conservative excision as done in the presenting case.The postoperative follow-ups did not show any sign of tumor recurrence.

VI. CONCLUSION

To conclude, the correct diagnosis of intraosseous myofibroma is crucial to prevent morbidity and to rule out other possible aggressive lesions as this can be corrected with conservative management. Myofibroma should be considered in the differential diagnoses of fibrous tumors with bone destruction in the orbit. The prognosis of this rare tumor is excellent even after conservative surgical curettage or resection.

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