Solitary Intraosseous Myofibroma of the Zygomatic Bone: A Case Report

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INTRODUCTION

Modified fibroblasts with smooth muscle like features (myofibroblasts) were first observed in granulation tissue of healing wound. The presence of myofibroblasts have been implicated in most fibro-contractive diseases and even in developing and specialized normal tissues.1

Lesions in this category fall into four main groups-reactive fasciitis-like lesions, benign lesions, the locally aggressive fibromatoses and, finally, sarcomas.2

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.3

It is a solitary nodular benign tumor of the soft tissue, bone, or internal organs that affects all ages. The tumor may present as single or multiple nodules.4

Intraosseous solitary myofibromas have been reported in both adults and children but are rare in adult jaws. The remaining intraosseous lesions have been reported in children.

This article describes the clinical, radiographic, histopathologic, and immunohistochemical features of an intraosseous myofibroma of the zygomatic bone in a female child.

CASE REPORT

A 7-year-old female presented with a swelling on left side of the face of 2 months duration. Swelling was insidious in onset, small to begin with and had gradually increased to attain the present size. There was no history of discharge, pain, fever, numbness or paraesthesia. There was no history of nasal discharge or pain on bending the head down. There was no history of change in voice. Visual acuity, extraocular motility, and pupillary functions were normal. Past dental, medical, family, and personal history were non-contributory. The patient was moderately built and well-nourished, and no gross physical or mental abnormalities were detected on doing thorough general physical examination.

A well-defined extraoral swelling, measuring approximately 1.2 cm in largest diameter was noted on the left zygomatic bone, situated at the inferolateral aspect of the left orbit. The

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overlying surface appeared normal with no surrounding erythema. The surface was intact with no secondary changes (Figure 1).

On palpation, the swelling was non-tender, with no local rise in temperature. It was bony hard in consistency, non-fluctuant, non-reducible, non-compressible and non-pulsatile. There was no bleeding or discharge evident on palpation with no audible/palpable bruit. Intra-oral examination ruled out any odontogenic etiology.

Computed tomography scan of peripheral nervous system and orbit revealed small (18 mm), lytic, expansile, fluid density lesion involving the left zygomatic bone, situated at the inferolateral aspect of left orbit. There was no intra-orbital extension of the lesion. Intra-orbital structures and rest of the bony orbital walls appeared normal (Figure 2).

The tentative clinical differential diagnosis included monostotic fibrous dysplasia, simple Bone cyst, central ossifying fibroma, giant cell tumor of bone, and aneurysmal bone cyst.

The lesion was surgically explored under GA. Left lower blepharoplasty incision was given. After blunt dissection in infraorbital region, an area of surface bony erosion was exposed (Figure 3). 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 (Figure 4).

Histopathology of the excised tissue showed that the tumor was composed of spindle cells with elongated nuclei and tapering to blunt ended nuclei and moderate eosinophilic cytoplasm arranged in fascicles and whorls. Stroma was fibrous and myxomatous with basophilic appearance in areas. Focal increased vascularity with hemangiopericytoma like the pattern was seen. Mitotic activity was not increased. Periphery showed lamellar bone rimmed by osteoblasts. There was no evidence of tuberculosis and malignancy. An impression of intraosseous myofibroma was given (Figure 5).

Immunohistochemical stains showed diffuse cytoplasmic reactivity for vimentin and focally positive Smooth Muscle Actin in the peripheral myoid cells, while negative in the central myofibroblastic cells (Figure 6).

A diagnosis of intraosseous myofibroma was established. The case was followed up every 3 months for a year for any evidence of recurrence or discomfort and the patient reported none (Figure 7).

**DISCUSSION**

Stout, in 1954 first reported congenital generalized fibromatosis. Tumor has been described under different nomenclatures in literature including generalized hamartomatosis, multiple congenital mesenchymal tumors, diffuse congenital fibromatosis, multiple vascular...
leiomyoma of the new born, infantile myofibromatosis, and myofibroma.4

Myofibroblasts have been implicated to play a major role in wound healing, therefore, it has been hypothesized that trauma or injury can be attributed to, at least partly, for their development.6 The theory has been supported by the fact that some adult myofibromas were stained positive with endothelial antigen BNH9 and CD34. Furthermore, it was put forward that myofibromas are a reactive vascular lesion, and the cell of origin could be a primitive vascular progenitor cell and differentiation into myofibroblasts, pericytes, and endothelial cells which are observed in this lesion.7

The etiology remains uncertain, although numerous theories have been put forward. Both autosomal dominant and autosomal recessive modes of inheritance have been suggested as seen by an increased occurrence within families and among twins.8 No family history was implicated in the present case. As the disease is most common seen to occur at a younger age, an intra-uterine toxic state during pregnancy has been implicated as an etiological factor. Exposure to estrogens in-utero has been suggested to be a potential contributing factor. Elevated levels of urinary basic fibroblast growth factor were reported in an infant with this tumor which suggested the role of angiogenic factors in its pathogenesis. Deletion of chromosome 6, (del [6] [q12q15]) was also noted in one of the cases, although there is not enough evidence in literature to support the theory.9

Myofibromas can be divided into two separate types; the solitary form and the multicentric form. The solitary form is defined by the presence of a single nodule on the skin, bone, muscle, or subcutaneous tissue; while the multicentric form is subdivided into further two types, the multiple form and the generalized form. The multiple form also involves bone, and the generalized form which also involves viscera.10 There is a higher incidence of the solitary myofibroma than the multiple type.4 Almost half of the cases of solitary myofibromas arise in the cutaneous/subcutaneous tissue of the head and neck region, followed by trunk, lower, and upper extremities and the other half of the cases are seen in skeletal muscle or aponeuroses, with a small number of cases involving bone, predominantly calvarial bone.11 The present case was a case of solitary myofibroma involving the calvarial bone in a female child although literature reveals a slight male predilection (about 62%) for both the solitary and the multicentric form. Both solitary and multicentric lesions can occur over an extremely wide age range that extends from new-borns to the elderly.10

As also seen in the present case, intraosseous myofibromas are often asymptomatic at the time of diagnosis, with a
swelling forming the initial presentation. On a cut section, myofibromas appear firm, fibrous with a gray-white, light tan to brown, or purplish surface. The central core is often yellow or necrotic or a cystic area filled with caseous-like material or hemorrhage may be noted.\textsuperscript{11}

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.\textsuperscript{12} Hyalin-like areas may be present and areas of less-differentiated cells arranged in a hemangiopericytoma-like pattern have been reported. The histopathology of the present case was highly characteristic. Areas of calcification have also been reported in literature which generally occur in lesions with necrosis, not evident in the present case.\textsuperscript{13}

The spindle cells of intraosseous myofibroma have the ultra-structural and immunohistochemical characteristics of myofibroblasts, staining positive for vimentin and smooth muscle actin, but negative for desmin.\textsuperscript{14} Cells are also negative for S-100 protein, allowing their differentiation from more immature histiocytes.\textsuperscript{9}

Treatment of myofibroma is usually conservative excision as done in the presenting case. In only a few cases, aggressive surgical procedure might be indicated if the tumor behavior is aggressive and destructive. Studies suggest that the recurrence rate is extremely low, therefore, it is suggested that the available potential anti-myofibroblast pharmacological therapeutic agents could be considered to spare these patients from aggressive surgical procedures. Selective estrogen receptor modulators, i.e., tamoxifen, raloxifene, and toremifene have been used with a fair degree of clinical success in another more aggressive myofibroblastic lesion, e.g., the fibromatoses.\textsuperscript{15} Their action on myofibroblasts is not dependent on the presence of estrogen receptors. It has been proposed that in the future, these agents, either alone or in combination with limited surgery, be used in cases of large and destructive myofibroma.

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. The correct histopathological and immunochemistry plays a pivotal role for correct diagnosis and therapeutics. The prognosis of this rare tumor is excellent even after conservative surgical curettage or resection.

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