Solitary Infantile Myofibromatosis at Distal end of Humerus: A Rare Case Report

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ABSTRACT

Introduction: Infantile myofibromatosis is a rare benign disorder of mesenchymal origin presenting as tumors in skin, muscles,bones and soft tissue of head and neck. Its solitary occurrence in bones is very rare. It is even rarer in peripheral location of skeletal system.

Case report: Herein we report a case of infantile myofibromatosis at distal end of humerus confirmed by histopathological examination. Patient was successfully treated with curettage and filling of cavity by bone substitute (beta tricalcium phosphate).

Conclusion: Infantile myofibromatosis is a rare event. Despite of its rarity a high index of suspicion must be practiced to arrive at correct diagnosis and treatment.

Keywords: Solitary, Infantile myofibromatosis, bones, beta tricalcium phosphate.

INTRODUCTION

Infantile myofibromatosis(IM), first described by Stout[1] in 1954, is characterized by benign myofibroblastic tumors in the soft tissues, the bones, and, occasionally, the viscera. The disorder can primarily be separated into two types although some texts classify it into three of four types as well.[2-5] The most commonly documented is the solitary form with a nodule in the skin, bone or viscera and the other is the multicentric type. Solitary skeletal lesions are relatively uncommon but when present, they occur more frequently in the craniofacial skeleton. The bones most commonly involved are skull, femur, tibia, spine or ribs.

CASE REPORT

We report the case of a three year old male child who presented with chief complains of stiffness and a painless swelling of size 2x2 cm over the posterior aspect of the right elbow along with stiffness. The swelling had been present for six weeks and was non tender and firm in consistency. No lymphadenopathy or visceromegaly was noted. The child had no other significant medical history. Patients family history was found to be insignificant.

An X-ray of the right elbow revealed a radiolucent, expansile lytic lesion over the medial epicondyle surrounded by dense sclerosis in the distal end of the humerus[Figure1]. Since all other clinical examination was found to be insignificant, the patient was not sent for further X-rays in lieu of radiation exposure and was kept on regular follow up. The prognosis was explained to the child's parents who concurred with our approach to his treatment.

The patient was admitted and the basic investigations done. On the 5th day after admission, the patient was taken into surgery and was managed by open biopsy, curettage and chronose granules (beta tricalcium phosphate) application[-Figure 2]. Above elbow plaster of paris slab was applied for 4 weeks. After 4 weeks active elbow range of motion physiotherapy was started. Patient regained full range of movements at elbow joint at the end of 6 months and x ray of elbow showed healed lesion [Figure 3]. Patient was followed for 18 months and there was no evidence of recurrence or any other complication.The biopsy sample was sent for histopathological examination and the reports confirmed it to be infantile fibromatosis.

DISCUSSION

IM is the most common fibrous tumor in early childhood, even though its overall incidence is low.[6] About 90% of these are found in patients less than 2 years of age and is rare in children and adults.[6] It is divided into two types: single solitary lesions as noticed in our case and multicentric type. The multicentric variant is further sub-divided into two types depending on whether there was any visceral involvement involving lungs, gastrointestinal tract (liver and pancreas) and kidneys.

Chung and Enzinger[7] and Muraoka[8] et al found the solitary lesion to be the most common mode of presentation (50-75%)that generally affects the skin, muscle, bone and subcutaneous tissue in the head, neck and trunk. The nodules are well-circumscribed, painless, firm and associated with

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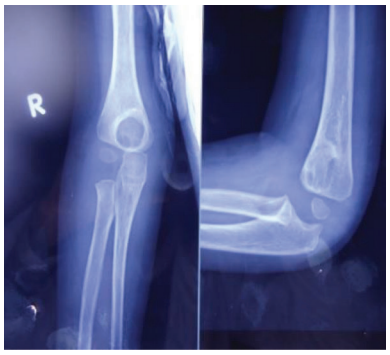

Figure-1: X ray of elbow showing a radioluscent, expansile lytic lesion in the medial epicondyle of humerus surrounded by dense sclerosis.

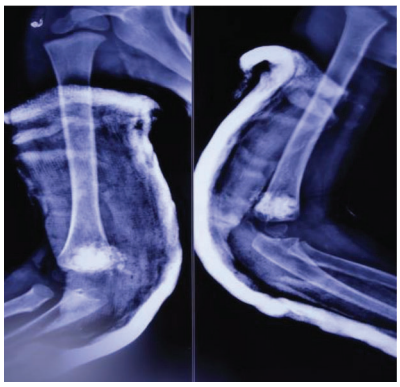

Figure-2: Post operative X ray of elbow showing post curretage cavity filled with bone substitute.

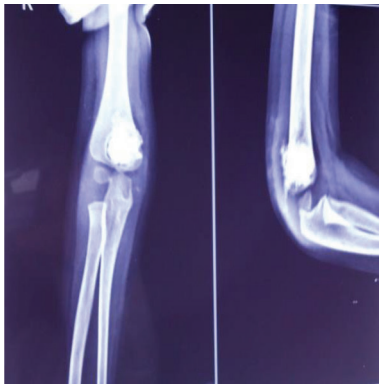

Figure-3: Six months follow up X ray of elbow showing healed lesion with incorporated bone substitute material.

an initial phase of rapid growth.[9] They are more commonly found in males

Besides the routine blood investigation, the suspicion of infantile myofibromatosis warrants the following investigations in all cases: x-rays of the long bones and skull, chest CT, abdominal and pelvic ultrasound and echogram. These investigations don't confirm the diagnosis but are a play a significant role in ascertaining the extent, progression and the chance of recurrence of the disease. The diagnosis is confirmed by biopsy as in our case. On histopathological examination, it is found to have ovoid to spindle-shaped collagen-forming cells showing immunohistochemical and electronic characteristics intermediate between fibroblasts and smooth muscle cells.

The patients with solitary lesions are usually treated conservatively unless it causes symptoms as it had in our patient. In such cases, an operative intervention can be planned as was done for our patient by open biopsy, curettage of the lesion and chornose granules application to fill the defect. If managed conservatively, regular follow ups are essential to monitor the progression.The multicentric variant has a poorer prognosis with the mortality of over 70% in patients with visceral involvement.

Treatment for the solitary form is expectant with clinical and imaging follow-up due to the possibility of spontaneous regression. The multicentric form requires a surgical approach. Surgical treatment is also considered appropriate when the tumor causes clinical compromise as happened with our patient who had respiratory failure due to tumor growth in the upper airway. The generalized variant of IM has a poor prognosis and use of CT may be considered. After conservative treatment (periodic evaluation to determine spontaneous regression) or surgical treatment, follow-up of these patients must be done because of the possibility of recurrence. This usually is ~5% for the solitary form. There is also the possibility of recurrence in the case of incomplete and inappropriate curettage or even after excision. Prognosis depends on mode of presentation. It is usually benign with spontaneous regression during a total period of 1-2 years for the solitary variant and also for the multifocal variant without visceral involvement. The multicentric form with GI, cardiac or pulmonary compromise may have a mortality rate of up to 73%.14

CONCLUSION

Infantile myofibromatosis is a rare myofibroblastic tumor usually seen in axial skeletal. Its solitary presentation in bone is even more rarer. Though rare it nevertheless warrants greater emphasis than it receives in literature. High index of suspicion should be practiced for correct diagnosis and treatment.

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