

NORA'S LESION-A RARE CASE REPORT**N. Lal¹, N. Tiwari¹, P. Gupta^{1*}, F. A. Siddiqui¹, S. Z. Rizvi¹, A. Jaiswal¹, O. Musa²**¹Department of Pathology, Era's Lucknow Medical College and Hospital.²Department of Surgery, Era's Lucknow Medical College and Hospital.Article Received on
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Corresponding Author*Dr. P. Gupta**Department of Pathology
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College and Hospital.**INTRODUCTION**

Fibro-osseous pseudo tumor is a rare benign lesion reported as a differential diagnosis of a painful calcified swelling in the finger. Ossifying lesions of soft tissue are fairly uncommon. However they may be a cause of concern to the clinicians as extra skeletal osteosarcoma an aggressive bone forming lesion is a differential in such cases ^[1]. These cases give a clinically aggressive appearance leading to radical treatment of the lesion when actually the lesion is benign.^[2,3] More than 45 cases of pseudo tumors have been reported till now^[4,5] and to the best of my knowledge this is the first case to be reported from India. Fibro-osseous pseudo tumor of the digit is

intriguing due to its odd location, i.e. tips of fingers and toes. However ours is the first case to be diagnosed in the thenar region. It is a superficially located lesion, in the dermis, therefore chances of ulceration and secondary infections are high. There is no racial or ethnic predilection and there is slight female preponderance and is thought to be a reactive process.^[6-8] Early diagnosis and resection of fibro-osseous pseudo tumor is important as it is a rare entity, having potential for misdiagnosis and overtreatment. Although till date no malignant transformation of fibro-osseous pseudo tumor has been reported, but a definite link has been seen between long standing chronic inflammatory or irritable lesions and neoplasia, therefore early excision is necessary. In difficult cases a punch biopsy can lead to a definite diagnosis.^[9]

CASE

Our case is that of a 30 year old Female, who had a large painful mass on the thenar aspect of her right palm for three years. There was no associated joint swelling, or redness but restriction of joint mobility was seen. The patient did Not recall any specific history of trauma. The biopsy revealed a dermal fibro-osseous lesion and therefore excision was

advised. The lesion measured 10x9x5 cm and no connection with underlying bone was demonstrated. However due to the size of the mass and the restriction in joint mobility her hand was amputated and sent to the Pathology department of Era's Luck now Medical College and Hospital.

Grossly the specimen consisted of the entire right hand with a large mass on the thenar aspect covered with skin measuring 10x9x5 cm. Microscopic sections examined revealed biphasic fibro-osseous lesion, composed of stromal and osseous components. Overlying skin appeared mildly hyperplastic and borders of the lesion were well circumscribed. Stromal component of the tumor is composed of bland spindle shaped cells with interspersed blood vessels. Stromal cells appear fibroblastic with elongated nuclei, eosinophilic cytoplasm and indistinct cell borders. They show minimal nuclear atypia and pleomorphism. Osseous component was formed by mature bony trabeculae with prominent osteoblastic rimming. Zonation pattern was not evident in the lesion. No mitotic activity was noted.

DISCUSSION

Fibro-osseous pseudo tumor of the digit is a rare benign ossifying lesion which has been called in the past by various names like florid reactive periostitis, parosteal fasciitis and fasciitis ossificans.^[10-12] It commonly affects young adults from 20- to 30-years-old, with women predominance.^[11] 40% to 50% of the cases are associated with trauma before development of lesion.^[13] Pathogenesis of fibro-osseous pseudo tumor is thought to be related to repeated trauma to the area.^[14]

Main pathologic differentials to this entity are myositis ossificans, subungual exostosis and extra skeletal osteosarcoma. Fibro-osseous pseudo tumor is closely related to myositis ossificans and some authors regard it as a cutaneous counterpart of myositis ossificans^[15]. However, myositis ossificans usually occur after trauma, in the deeper aspect of proximal soft tissues, and histopathologically show a typical zonation pattern.^[16] Subungual exostosis may clinically simulate fibro-osseous pseudo tumor, but radiologic connection to the underlying bone and histopathologic demonstration of fibrocollagenous cap separate out the two entities.^[17] Finally extra skeletal osteosarcoma should always be ruled out; however it shows destructive stromal invasion, obvious cytologic atypia and immature osteoid directly formed by tumor cells.^[18]

These tumors are seen to be positive for Anti smooth muscle actin ASMA.^[19]

In diagnosing such a tumor it is beneficial to follow a pattern for e.g.; stromal component should be evaluated first, for evidence of malignancy including nuclear atypia, pleomorphism, mitotic activity and growth pattern. If the stromal component of the tumor appear malignant, then the differentials would include osteosarcoma and other dedifferentiated sarcomas with heterologous differentiation.

If the stroma appear fibroblastic then osseous component should be further uncovered for maturation and osteoblastic rimming. With mature osteoid and prominent osteoblastic rimming, the main differential would be fibro-osseous pseudo tumor and myositis ossificans. Distal and superficial locations with absence of zonation pattern favor the diagnosis of fibro-osseous pseudo tumor while deep lesions in proximal location and zonation pattern will suggest the diagnosis of myositis ossificans.^[9]

Clinically it presents as a rapidly enlarging painful mass, typically adjacent to one of the phalanges. It features heterotopic or met aplastic ossification and is closely related to myositis ossificans. Malignancies and acute fulminating supportive tenosynovitis are important differential diagnoses. Unlike classical myositis ossificans, which chiefly affects the muscle fibers, this lesion mainly arises from the sub cutis and adjacent fibrous tissue. It lacks the well-defined zoning pattern seen in myositis ossificans ^[1] It can be differentiated from acute calcium deposition in fingers by a well-defined ossification process, which sometimes has features of chondroid tissue formation. Florid reactive periostitis of the tubular bones of the hands and feet and parosteal fasciitis are identical lesions.

Fibro-osseous pseudotumour has also been reported to present as a subungual tumour. Histologically, it also resembles bizarre parosteal osteochondromatous proliferations (Nora's lesion). Characteristic of a Nora's lesion is its attachment to the underlying bone, arising from the cortical surface without disturbing its medullary architecture. Early and radical excision of this lesion is crucial in preventing painful recurrence. ^[19] Prognosis of fibro-osseous pseudo tumor is good with complete excision being curative without evidence of local recurrence ^[3, 9].



FIGURE A-Gross image of tumor involving the thenar area of right palm.

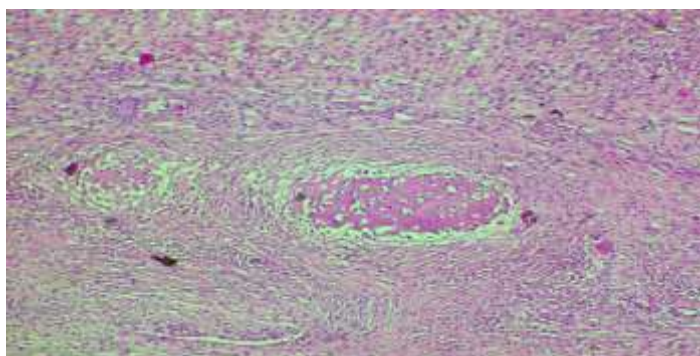


FIGURE 1-Microscopy of spindle cells with mature cartilage[H&E;100X]

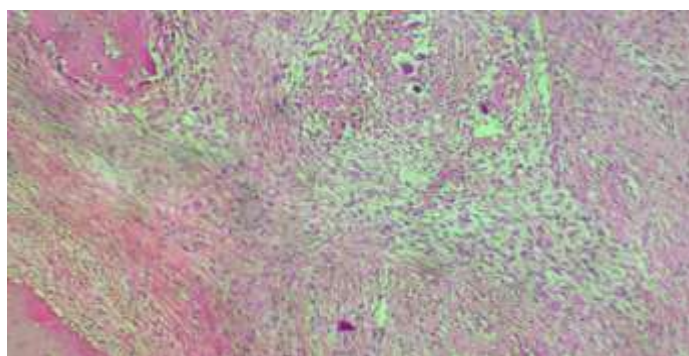


FIGURE 2-Microscopy showing benign spindle cells with mature cartilage and few giant cells [H&E;100X]

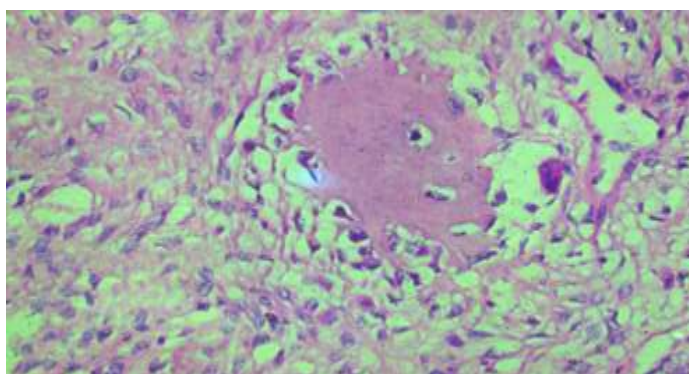


FIGURE 3- Showing cartilage island with a giant cell in fibrous tissue [H&E;400X]

CONCLUSION

Fibro-osseous pseudo tumor is a rare benign reactive condition which can mimic many malignant lesions so its early diagnosis and treatment is important.

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