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A RARE CASE OF FIBROUS HAMARTOMA OF INFANCY

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ABSTRACT

Subcutaneous soft tissue lesions in infants and children encompass a diverse spectrum of entities including benign and malignant lesions. The rarity and diversity of these lesions presents diagnostic and therapeutic challenges. Fibrous hamartoma of infancy (FHI) is an uncommon, benign tumour of subcutis and lower dermis. More than 90% of the lesions occur during the first year and approximately 25% are congenital. The lesion is more common in males and the sites include axillae, chest wall, back, thigh, and the inguinoscrotal region. The clinical and radiologic features of this lesion are non-specific and can mimic those of malignant soft tissue tumours. Diagnosis is made by histologic examination to identify the characteristic triphasic pattern arranged in an organoid fashion. It has an excellent prognosis with 16% local recurrence rate. Hence, we report this case of an 8-month-old male child presenting with a swelling over the lateral side of chest wall for 3 months which was progressively increasing.

KEYWORDS: infancy, subcutaneous, fibrous.

INTRODUCTION

Fibrous hamartoma of infancy (FHI) is a rare, benign tumour of subcutis and lower dermis, which usually occurs within the first 2 years of life.[1] These lesions are solitary and present most commonly over the axillae, arm, shoulder, chest wall and back.[2] These lesions belong to a genre of their own and are quite liable to be misdiagnosed as a malignant soft tissue neoplasm of infancy and risk being overtreated with catastrophic results. Diagnosis is made by histologic examination to identify the characteristic triphasic pattern.[3]

CASE REPORT

An 8-month-old boy presented with a soft, swelling in the left side of the chest wall. His mother had noticed this lesion 3 months back which was progressively increasing in size. Physical examination revealed a firm solitary, non tender, mobile mass in the left side of the chest wall measuring 5x4 cms. The lesion was in the subcutaneous plane with no attachment to the underlying muscle or bone. Overlying skin was normal. There were no similar lumps anywhere over the body and no lymphadenopathy. A clinical suspicion of lipoma was done. FNAC was attempted which showed only hemorrhage.

After complete hematological, biochemical and pre anaesthetic workup the child was posted for surgical excision of the lesion. The mass was excised and sent for histopathological examination. We received a well encapsulated firm mass measuring 5x4cms, greyish white with no areas of hemorrhage. Microscopy showed interlacing trabeculae of fibrocollagenous tissue, mature adipose tissue [fig 1, fig 2] and loose textured areas of immature appearing spindle shaped cells [fig 3]. IHC was done with vimentin, which was positive [fig4]. Hence, a final diagnosis of fibrous hamartoma of infancy was given.

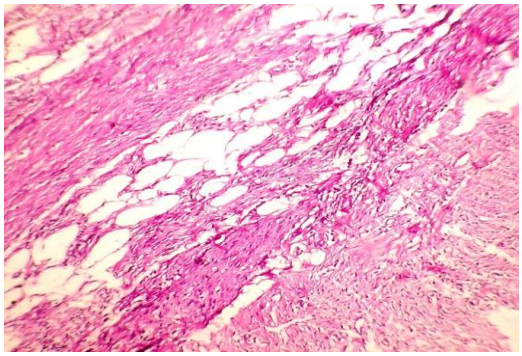

Fig 1: Showing mature adipose tissue interspersed between the fibrocollagenous tissue and areas of immature spindle cells (H&E, 10X).

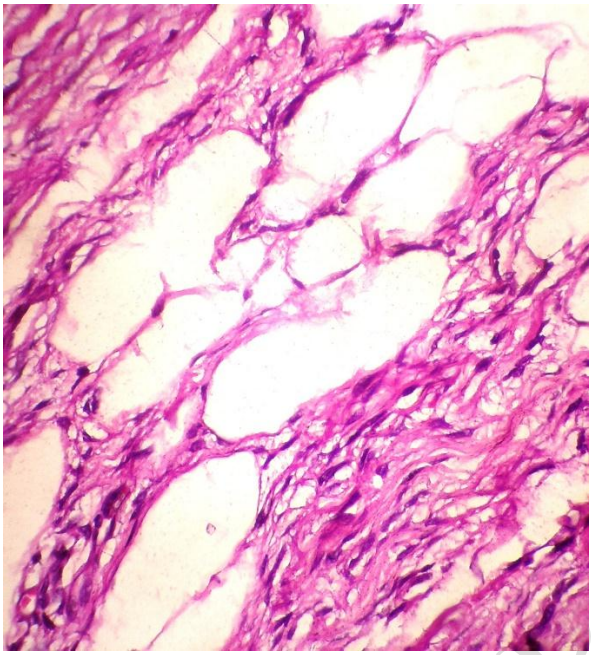

Fig. 2: areas showing adipose tissue and fibrocollagenous tissue (H&E, 40X)

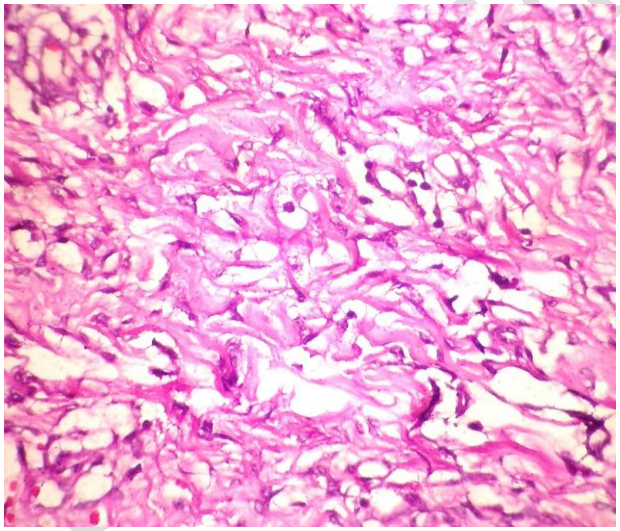

Fig. 3: immature spindle cells seen (H&E, 40X)

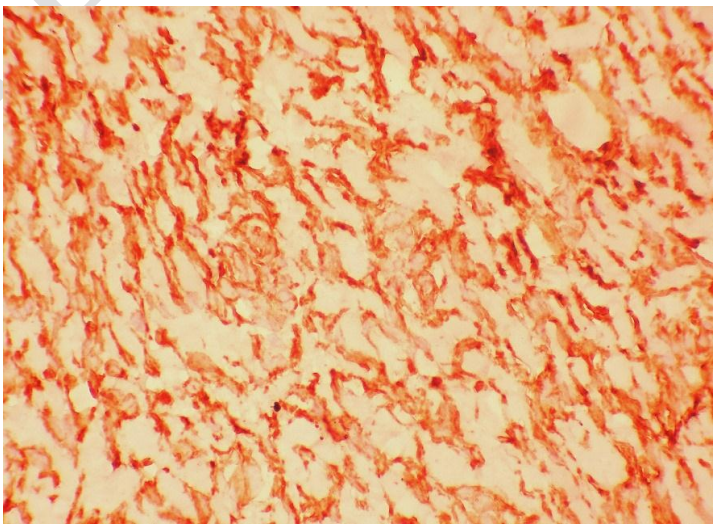

Fig. 4: vimentin positivity by IHC.

DISCUSSION

Some varieties of soft tissue lesions are noted among children, which have no clinical or morphologic counterpart in adults. FHI is one of these lesions with unique clinicopathologic features. It is an uncommon benign fibroblastic and myofibroblastic proliferation that typically occurs within the first two years of life. More than 90% of reported cases present during the first year of life, and 25 % of the cases have been described to be congenital.[4]

This rare entity was first described by Reye in 1956 and he called it 'subdermal fibrous tumor of childhood'. Enzinger in 1965 after a review of 30 cases nomenclatured the lesion as 'Fibrous Hamartoma of Infancy.[1] Till date very few cases have been reported in medical literature.[5] The size ranges from 0.5 cm to very large masses over 20 cm.[3] There is a distinctive male preponderance and the males being afflicted twice to thrice as more commonly over females.[6] Most cases present as solitary masses, but multiple separate synchronous lesions have been reported rarely.[3] Preoperative diagnosis by fine-needle has been established only in few cases.[7] Thus for definitive diagnosis, we need to have a complete excision biopsy

Grossly, it is poorly circumscribed with firm gray-white tissue admixed with fat. But in our case, it was well circumscribed. FHI has a specific histologic appearance comprising of three different mesenchymal tissues in variable proportions arranged in an organoid fashion: intersecting trabeculae of fibrocollagenous tissue, mature adipose tissue, and loosely textured areas consisting of immature small rounded to stellate cells embedded in a myxoid matrix. The spindle cells react primarily with vimentin and to a lesser extent with smooth muscle actin. Ultrastructural examination reveals an admixture of fibroblastic and myofibroblastic cells.[8]

Pediatric soft tissue tumors composed of adipose tissue and fibroblastic elements have been recently classified as lipofibromatosis by Fetsch et al. [9] Histologically they are composed of abundant adipose tissue traversed by bundles of fibroblast-like cells but there are no primitive mesenchymal cells as FHI. For differentiating FHI from infantile digital fibroma sites of involvement and presence of intracytoplasmic perinuclear inclusions can be useful. Myofibroma has a characteristic hemangiopericytoma-like pattern and calcifying aponeurotic fibroma has a zonation pattern which can be helpful in differentiating these lesions from FHI.[8,10]

Lakshminarayanan R et al, reported a reciprocal translocation, t (2; 3) (q31; q2) and a possible genomic loci on chromosome 2q31, coding the vitronectin receptor-α subunit of the integrin family.[10] The usually observed manifestation of FHI on CT and/or MR imaging is the strands of adipose/fibrous intensities traversing the lesions, with the characteristic parallel or whirling appearance in some cases. The tumours with ill-defined margins have the tendency to involve the underlying muscles.[11]

Carretto et al, in his evaluation of 18 cases of Fibrous hamartoma of infancy, concluded that treatment of choice for Fibrous hamartoma of infancy is wide local excision taking judicious margins with tissue reconstruction; and it is imperative not to over diagnose and over treat these benign lesions with radical and ablative procedures.[12] Although as many as 16% locally recur within a few months after primary excision, recurrence is non-destructive and cured by re-excision.[9]

CONCLUSION

Fibrous hamartoma of infancy is a rare benign lesion with typical cytomorphological features. A strong clinical acumen coupled with histopathological confirmation and subsequent prompt wide local excision is in the best interest of the patient.

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