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# Fibrous Hamartoma Of Infancy : A Case Report

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***Abstract***
***Introduction:*** *Fibrous hamartoma of infancy is a benign neoplasm of infant and young children showing characteristic triphasic histomorphology. Most of the cases of fibrous hamartoma of infancy (FHI) occur in young boys, aged < 2 years (M:F - 2.7:1). It is a rare tumor having 3 characteristic tissue components in variable proportions i.e. mature fibrous tissue, mature adipocytes and immature mesenchymal component. Clinically it may present as a solitary subcutaneous poorly delimited tumour. Occasionally, it may recur as is evident in the current case.*
***Case report:*** *Present patient was a male infant, aged 11 months. He had complaints of recurrence of a tumour on left lower thigh. It measured 4×3×1 cm. Tumour was excised and two pieces were collected. Both the pieces showed lobules of mature adipocytes with traversing bands of fibrous tissue of variable thickness, consisting of spindle cells. Loosely textured areas, containing ovoid to stellate cells in a myxoid stroma were also seen. Fascicular component may appear myofibroblastic. Areas of mononuclear inflammatory infiltrate were also noted. Myxoid areas may appear primitive. There is no mitosis. No areas of necrosis. Tumour was biopsied twice within a gap of 10 months due to local recurrence. IHC was subsequently performed for confirmation of the diagnosis.*
***Discussion:*** *FHI is a soft tissue tumour of subcutis and lower reticular dermis. Tumour may be asymptomatic with excellent prognosis. Treatment of choice is local excision. First biopsy was done 10 months earlier. Later, the patient developed recurrence. However, FHI has a low recurrence rate (~13%).*
***Conclusion:*** *Current lesion was a rare benign tumour, located at left lower thigh in a 11 months old male infant with history of recurrence. Surgical resection of tumour was done. Histological examination of tumour showed intermixed trabeculae of fibrous tissue and adipose tissue with interspersed islands of immature mesenchymal tissue in a myxoid stroma. Tumour was poorly delimited from adjoining tissue. The patient was diagnosed as fibrous hamartoma of infancy.*
***Keywords:*** *Spindle cells, adipocytes, myxomatous tissue, benign, asymptomatic.*

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## I. Introduction

First case of fibrous hamartoma of infancy (FHI) was described by Reye in the year 1956 as subdermal fibromatous tumour of infancy[1]. Later, Enzinger studied 30 cases and renamed it as fibrous hamartoma of infancy in the year 1965[2]. Most of these cases (91%) were reported during 1st year of life. Upto the year 2020 about 200 cases of FHI were reported. Rarely, a few cases (5%) have been described in adults[3]. A few FHI may begin in infancy and become symptomatic later in adult hood. Collection of excess amount of normal tissue, presenting as overgrowth or tumour may be called as hamartoma. It is assumed to be a disorganized embryonic maturation of normal cells. Often, the cells of hamartoma are irregularly differentiated[4]. Clinically, FHI may be silent and benign[5]. Further, triphasic morphology has been described for diagnosis of FHI, i.e. fibrous tissue, mature adipocytes and immature mesenchyme[6]. Commonly, FHI may present as a solitary subcutaneous tumour[6]. Approximately 25% of cases are congenital. Herein, we describe the features of a case of FHI.

## II. Case Report:

Current patient is an 11 month old male infant and had complaints of mass in the left lower thigh approximately 5cm in size with irregular margins. FNAC examination of the tumor was suggestive of spindle cell lipoma. The tumor was excised and sent for histological examination, the findings of which were suggestive of lipoma. Subsequently tumour recurred after 10 months of previous biopsy. Recurred tumour measured 4×3×2 cm. As per the discussion with the operating surgeon, intra-operatively, the tumour was a hard mass densely adhered with overlying skin and scar and was anterior and lateral to the upper end of tibia, fibula, knee capsule and interosseous membrane. Tumour was excised and two pieces were collected. Both pieces were sectioned. On histopathological examination, sections showed predominantly mature adipose tissue, areas of myxoid stroma with round to stellate cells and mature fibrous tissue suggesting three tissue components present in a soft tissue tumour in a variable proportion. Based on histo-morphology, an initial differential diagnosis of spindle cell lipoma and fibrous hamartoma was suggested. Histochemical stains were performed with Van Geisson stain which showed strong positivity with reddish collagen fibres. Masson's Trichrome stain showed strong positivity with fibro-collagenous tissue staining blue.

Subsequently, immunohistochemistry was performed with CD34, RB1, VIMETIN, SMA, BCL2, Ki-67 and S-100. The tumour showed strong diffuse immunoreactivity with anti-vimentin antibody. A positive reaction was also obtained with antiCD34 antibody which stained the vascular endothelial lining as well as the immature round to stellate cells embedded in a myxoid background. Both of the above proved mesenchymal origin of one of the tumour components. Anti S-100 antibody stained positive in adipocytic areas only. Anti smooth muscle actin antibody showed strong positivity in the fibrous tissue staining the nuclei of the tumor cells. Anti bcl2 antibody, on a contrary note, failed to stain the tumour cells. Negative reaction was also obtained with antiKi-67 antibody suggesting poor proliferative activity. Anti RB1 antibody also did not stain the tumor cells. The tumour was finally diagnosed to be fibrous hamartoma of infancy(FHI).

## III. Discussion:

In cases of FHI, tumour may be poorly delimited and usually may measure between 1 to 5 cm. Tumour shows sparse to no mitotic activity. Histologically, tumour may show triphasic morphology of mature adipose tissue, mature fibrous tissue and less-differentiated primitve mesenchymal cells [7]. According to meta-analysis study by L. Martos Cabrera et al, in cases of FHI, relative proportion of different tissue components is highly variable. A component is considered predominant if it occupies 45% or more of the entire lesion. FHI may present at birth or develop in the first 2 years life[1,8]. Rarely, it may occur in adolescence and adults but reports suggest the FHI manifesting at an adult age corresponds to a late diagnosis of the tumour rather than a late age of onset. Hamartoma in infants have been rarely reported. Few cases of congenital hamartomas have been described in infants. The patient is born with the tumour. It is believed to be a disordered embryonic differentiation of normal cells. It appears to be an overgrowth of one or more mature cells and one type of cells may predominate. Often cells of a hamartoma are irregularly arranged [4]. Pulmonary hamartomas are frequently reported at the age of ~60 years and are more common in females as compared with males. Frequently, they are located at the periphery, clinically silent and benign[5]. FHI may occur in axillary soft tissue and proximal parts of extremities[2]. Occasionally it may be seen adherent to the underlying fascia or insinuating to the surrounding intermuscular plane with extremely rare occurrence of muscle invasion and bony erosion noted in the cases of FHI. CT examination has a very limited specificity and plays only a supplemental role in the diagnosis of this lesion. On the other hand, MRI studies have a promising role due to high tissue resolution, contrast studies and diffuse weighted imaging and can help effectively distinguish between benign and malignant lesions. Histogenesis of FHI is still unclear. However, [t (2,3) q3:q21] reciprocal translocation is seen in cases of FHI[9]. In addition, an important gene 2q31 has been reported which supports the growth of FHI[9]**.** EGFR exon insertion/duplication mutations have also been noted in the pathophysiology of the tumour. Additionally, FHI may have low recurrence rate (~13%); probably recurrence occurs following inadequate surgical excision[10]. To prevent recurrence of the tumour, it is recommended to resect a clear surgical margin of at least 1cm with the depth of resection reaching the adjacent normal tissue. In cases of FHI with bony destruction, tumour resection with clear surgical margin of 1.5cm has shown a better result and a lower recurrence rate. Rarely, sarcomatous differentiation may occur in mesenchymal component of FHI. High grade areas have been described only in 2 of 145 cases of FHI[11]. In another report, 2 cases with sarcomatous change were described[12]. However, current case had all the 3 benign components of the tumour.

## IV. Conclusion:

Fibrous hamartoma of infancy is a benign neoplasm formed due to embryonic overgrowth of three tissue components i.e. adipocytes, fibroblasts and immature mesenchyme. Tumour may infiltrate the surrounding structures and may produce cell-death by necrosis and cause adjacent soft tissue and bone destruction. Rarely, mesenchymal component may get transformed into malignancy and present as a sarcoma. In conclusion, in an infant or a young child presenting with a subcutaneous soft tissue mass, FHI should be considered a significant differential diagnosis and investigations should be carried out to differentiate it from other differentials like lipofibromatosis, lipoblastoma, desmoids type fibromatosis, myofibroma, calcifying aponeurotic fibroma and giant cell fibroblastoma. FHI should also be differentiated from malignant neoplasms to avoid misdiagnosis and aggressive treatment. Local surgical excision with 1 cm clear margin in localized lesions and 1.5cm clear margin excision in lesion with bony destruction and unclear boundaries is recommended to avoid recurrence of the tumour.

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**Legends to figures**

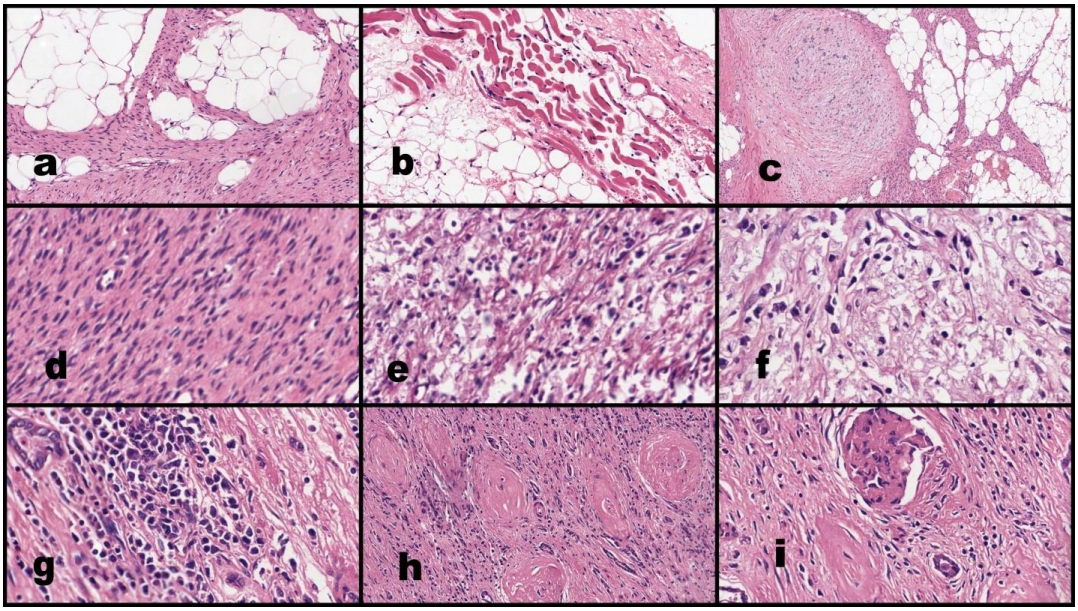


**Figure 1 (a) Photomicrograph shows bands of fibrous tissue traversing the lobules of mature adipocytes. Tumour cells had elongated nuclei. No mitoses seen(HEx200). (b)Photomicrograph shows mature adipocytes, skeletal muscle fibres showing splaying and area of necrosis(HEx200). (c) Photomicrograph shows tumour with myxoid stroma and islands of mature fibro-adipose tissue(HEx200). (d) Photomicrograph shows hypercellular tumour. Tumour cells have elongated, fusiform to spindle shaped nuclei. No mitoses seen in the tumour stroma(HEx400). (e) Tumour cells show primitive mesenchymal tissue, round to stellate cells in a myxoid stroma(HEx400). (f) Nuclei of tumour cells show mild anisonucleosis and myxoid stroma(HEx400). (g) Photomicrograph shows mononuclear lympho-plasmacytic inflammatory infiltrate arrange in cords and scattered diffusely in the tumour stroma and blood vessels(HEx400). (h) Photomicrograph shows immature mesenchymal tissue, numerous thin walled blood vessels and hyalinised blood vessels(HEx200). (i) Photomicrograph shows tumour stroma with hylanised blood vessel and sclerosed blood vessel in glomeruloid pattern(HEx400).**

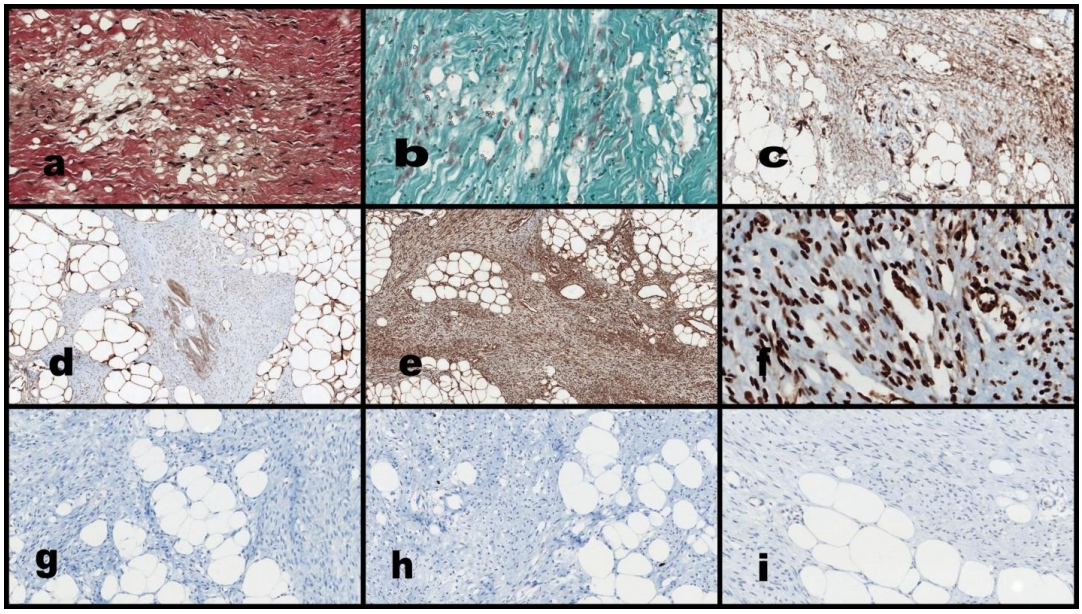


**Figure 2 (a) shows Van Geison stain with reddish collagen fibres and adipose tissue(VGx200). (b) Fibro-collagenous tissue staining blue with Masson's Trichrome(MTx400). (c) Anti CD34 antibody staining positive the primitive mesenchymal tissue and vascular lining endothelium. (d) Anti S-100 antibody stained positive only the lipomatous areas. (e) Anti Vimentin antibody gave a strong diffuse positive reaction (3+) in the tumour cells. (f) Anti SMA antibody strongly stained the fibroblast nuclei. (g) Anti Bcl2 antibody failed to stain the tumour cells. (h) Negative reaction was obtained with anti Ki-67 antibody suggesting poor proliferative activity (index ~5). (i) Anti RB1 antibody did not stain the tumour cells.**

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