Fibrous Hamartoma of Infancy in An Unusual Location- A Case Report

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Abstract

Fibrous hamartoma of infancy is an uncommon fibroproliferative lesion that occurs only in infancy and childhood. It predominantly affects healthy boys and can be found in almost any subcutaneous tissue. The tumor is most frequently found in the axilla, followed by shoulder, inguinal area, and chest wall. This tumor can cause much concern about malignancy because it is firm and may be fixed to underlying tissues. Despite the occasional local recurrence, the clinical course is benign and the prognosis is excellent. Treatment is by local excision. The diagnosis is made by the characteristic histologic features. In order to avoid the misdiagnosis of malignancy and unnecessary radical therapy, both surgeon and pathologist must be familiar with this entity. We report a case of fibrous hamartoma of infancy in a 4-year-old boy in gluteal region.

Key words

Fibrous hamartoma of infancy, myofibroblast, organoid

Introduction

Fibrous hamartomas of infancy (FHI) are uncommon tumors, accounting for less than 2% of soft tissue tumors occurring in the first year of life and approximately 5% of fibroproliferative tumors in children[1-2]. The tumor is an uncommon lesion of uncertain histogenesis. The lesion is benign, found mainly in boys younger than 2 years of age and can be seen at birth. The tumor arises from the subcutaneous tissue and is most frequently found in the axilla, followed by the shoulder, inguinal area, and chest wall. Because it is firm, it may be fixed to underlying tissues and exhibits high degree of cellularity along with presence of immature cells, it can be erroneously diagnosed as malignant. The treatment of choice is local excision. We report a case of fibrous hamartoma of infancy in child's gluteal region, an unusual location.

Case History

A 3-year old boy presented to our institute with the soft tissue swelling of left gluteal region of one year duration. The child was unable to sit in squatting position. There was no family history of similar skin or subcutaneous lesions. It started as a small peanut size swelling and gradually increased to 3x2 cms. On examination the mass was firm, non-mobile and had ill-defined borders. There was no evidence of inflammation. Radiographs revealed a soft tissue mass with ill-defined margins and without direct infiltration of bony structures. On FNAC benign fibroblastic lesion was considered and excision biopsy was advised. Grossly, the excision biopsy was flat, smooth, skin covered tissue piece measuring 3.5×3.5 cm in size. Cut sections revealed a glistening gray-white fibrotic appearance with focal yellow areas resembling fat. On histopathologic examination, there were multiple foci of immature organoid arrangements of primitive mesenchymal cells. In addition, the soft tissue mass revealed cellular areas composed of spindle shaped cells in fascicles along with focally entrapped mature adipose tissue (Figure 1). The lesion was mainly confined to the dermis and focal infiltration of underlying skeletal muscle was also noted (Figure 2). However, nuclear atypia or mitotic figures were not found. The histologic diagnosis of fibrous hamartoma of infancy was given.

Discussion

Fibrous hamartoma of infancy (FHI) is a rare, benign, fibroproliferative soft tissue tumor that usually occurs within the first two years of life. Fibrous hamartoma was first described by Reye

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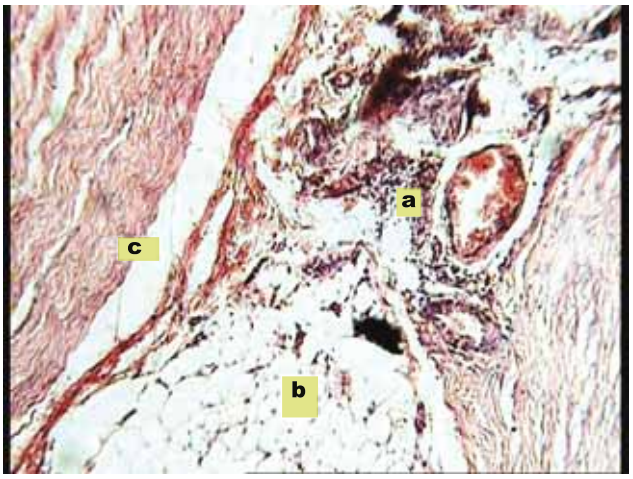

Fig. 1 : The microphotograph shows (a) small nodular aggregate of immature mesenchyme, (b) mature adipose tissue, (c) fascicular fibroblastic component (H&E, X100)

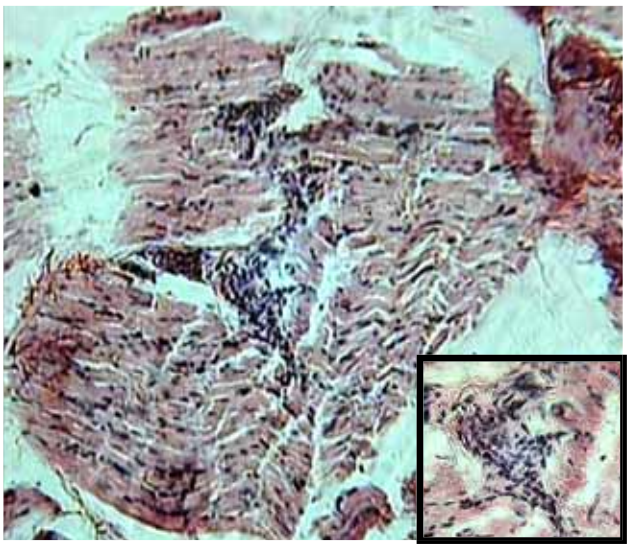

Fig. 2 : The microphotograph shows muscle infiltration by primitive cells (H&E, X100). The inset shows high power view of the same (H&E, X400)

in 1956 as a subdermal fibromatous tumor of infancy[3]. The term fibrous hamartoma of infancy was later coined by Enzinger in 1965[2]. Males are affected more often compared to females. Its histogenesis is also unclear. Fibrous hamartoma of infancy is most commonly found in the axilla, shoulder, upper arm, inguinal region, and chest wall; however, isolated cases have been reported involving the foot, scalp, perianal region, gluteal region, and scrotum[2-4] . It is usually a solitary malformation located in the subcutaneous tissue or reticular dermis. The lesions are typically 1 to 8 cm in diameter but have been reported up to 10 cm[4]. The tumor is usually firm, may be affixed to underlying tissue and sometimes exhibit high degree of cellularity, thus causing concern of potential malignancy[4]. Local recurrence is uncommon and treatment is largely successful by local excision. The clinical course is typically benign and prognosis excellent[4].

The characteristic histologic features of FHI show organoid pattern including admixture of well-differentiated spindle cells comprising of fibroblasts and myofibroblasts, accompanied by deposition of collagen, regions of mature adipose, and scattered primitive mesenchymal cells[3]. Anaplasia is not seen and mitotic figures are uncommon[5]. Ultrastructural studies have demonstrated the composition of FHI to include fibroblasts, myofibroblasts, primitive mesenchymal cells, small blood vessels, and mature adipocytes[6]. Immunohistochemical studies of FHI support the ultrastructural findings[6].

Local excision is the treatment of choice for FHI. The management of a subcutaneous mass in the pediatric patient presents a clinical challenge. The differential diagnosis includes both benign and malignant soft tissue tumors. A number of entities are included within the differential diagnosis such as epidermoid cyst, recurring digital fibrous tumor, juvenile aponeurotic fibroma, juvenile hyaline fibromatosis, palmoplantar fibromatosis, histiocytoma, dermatofibroma, leiomyosarcoma, and fibrosarcoma[7].

In conclusion, the physical appearance and characteristics of a subcutaneous mass on a child may suggest a malignant process; however, FHI should be included within the differential diagnosis. Furthermore, the prognosis of such diagnosis is excellent and local surgical excision is effective with rare events of recurrence.

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