Fibrous hamartoma of infancy

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ABSTRACT

Fibrous hamartoma of infancy (FHI) is a rare, benign tumor of the subcutis and lower dermis, which usually occurs within the first 2 years of life. Ninety one percent of the tumors occur in the first year of life. The histogenesis of FHI is unclear. The clinical course is typically benign and prognosis excellent. The physical characteristics of the subcutaneous mass in a child may suggest a malignant process; however, FHI should be included in the differential diagnosis. The prognosis of FHI is excellent with local surgical excision and it rarely recurs.

Key words: Fibrous hamartoma of infancy, pediatric soft tissue tumors

INTRODUCTION

Fibrous hamartoma of infancy (FHI) is a rare, benign tumor of the subcutis and lower dermis, which usually occurs within the first 2 years of life.[1,2] Ninety one percent of the tumors occur in the first year of life. The histogenesis of FHI is unclear. The clinical course is typically benign and prognosis excellent. We describe a 1.5-year-old female child with a mass on her left thigh that progressively increased in size.

CASE REPORT

A 1.5-year-old female child was brought to the pediatric oncology department for evaluation of a residual mass on her left thigh. The mass had been present for several months and had progressively increased in size. The child had undergone excision of the swelling 2 months back in a remote area and there was no histopathological report or slides for review.

On physical examination, an 8 cm scar of the previous surgery was seen on the back of the left thigh. A diffuse mass was felt underneath the scar with indistinct borders. There was no tenderness, erythema, or warmth associated with the mass. Range of motion, motor and sensory examination of the left leg was normal. Rest of the systemic examination was normal.

Radiographic analysis of the mass revealed no bony pathology. Magnetic resonance imaging (MRI) of the thigh revealed a diffuse soft tissue swelling with postoperative changes.

Re-excision of the mass was planned. Because of the previous surgery, the residual mass was adherent to the scar. Complete excision of the residual mass and the previous surgical scar was done. The remaining skin defect was too large for the primary closure and hence a split skin graft was placed [Figure 1].

Gross examination of the mass revealed poorly circumscribed, lobulated fragments of yellowish tissue measuring 2 × 1.5 × 0.5 cm. The mass was composed predominantly of whitish tissue of fibrous appearance intermixed with fat. Microscopically, the lesion showed a distinct organoid pattern, involving the dermis and subcutaneous fat. The lesion showed three distinct types of tissues: well differentiated fibrous tissue, mature adipose tissue and immature cellular area arranged in a whorl like pattern [Figure 2], typical of fibrous hamartoma of infancy. There was superficial involvement of the underlying muscle fibers. Base of resection and surgical margins were free of the tumor.

The child is asymptomatic after 1 year of follow-up with a well-healed wound.

DISCUSSION

FHI was first described by Reye in 1954.[1] Enzinger studied 30 cases and coined the term fibrous hamartoma of infancy.[2] It is a rare, benign

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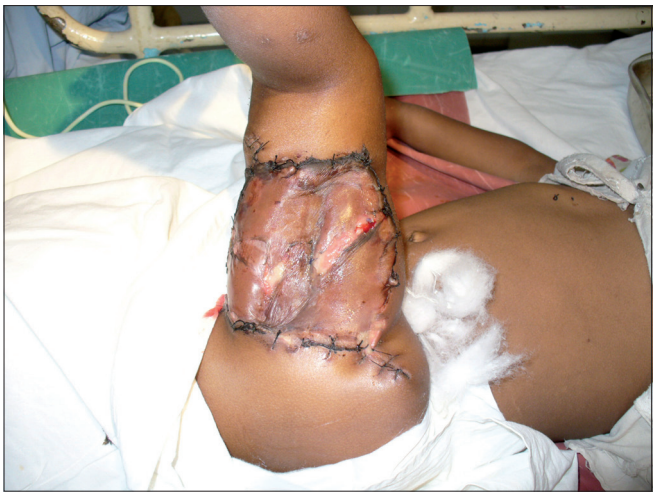

Figure 1: Skin defect after the excision is covered with split skin graft

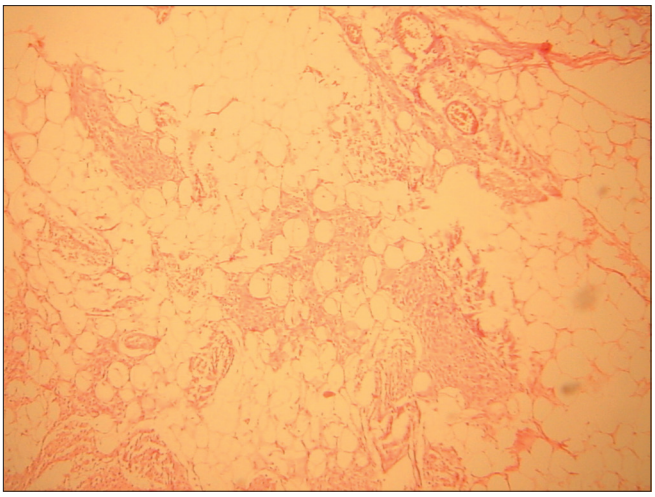

Figure 2: FHI shows an organoid pattern and is composed predominantly of mature adipose tissue, fibrous trabaculae and spindle-shaped cells (H & E, ×10)

tumor of subcutis and lower dermis, which usually occurs within the first 2 years of life. The vast majority of these cases occur within the first year of life (91%). Twenty-three percent are congenital. There is a predilection for boys with a male/female ratio of 2.4:1. FHI 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, perineal region, gluteal region and scrotum.[3-5] Thigh is an unusual site of presentation. Survey of the available literature revealed two reported cases of FHI in the thigh.

It is usually a solitary malformation located in the subcutaneous tissue or reticular dermis. The tumor is usually firm and may be affixed to underlying tissue, thus causing concern of potential malignancy. Local recurrence is uncommon and treatment is largely successful by local excision. The clinical course is typically benign and prognosis excellent.[3]

These tumors on gross examination, are round or bosselated and on section consist of firm grayish white tissue intermixed with irregular small islands of yellow fat. Most tumors blend partly or completely with the surrounding subcutaneous fat. The lesions are typically 1–8 cm in diameter but have been reported up to 10 cm.[6,7] Microscopically, three distinct kinds of tissues could be identified (1) well-defined traversing bundles or trabeculae of dense fibrocollagenous tissue somewhat resembling fetal tendons; (2) immature-appearing, loose textured cellular areas; and (3) mature fat interposed between the other two components.[2] Ultrastructural studies have demonstrated the composition of FHI to include fibroblasts, myofibroblasts, primitive mesenchymal cells, small blood vessels, and mature adipocytes.[4,8] Immunohistochemical studies of FHI support the ultrastructural findings.[4,9]

Local recurrence rate is approximately 16% according to a study by Enzinger and may have been explained by incomplete excision of the lesion.[2] The clinical course of FHI is benign but untreated cases continue to grow without regression. Local excision is the treatment of choice for FHI; however, delayed surgery is not associated with an increased risk of operative complications.[6]

The differential diagnosis for subcutaneous swellings in an infant includes both benign and malignant soft tissue tumors such as epidermoid cyst, recurring digital fibrous tumor, juvenile aponeurotic fibroma, juvenile hyaline fibromatosis, palmoplantar fibromatosis, histiocytoma, dermatofibroma, leiomyosarcoma and fibrosarcoma.[10]

In conclusion, the physical characteristics of a subcutaneous mass in a child may suggest a malignant process; however, FHI should be included in the differential diagnosis. The prognosis of FHI is excellent. It rarely recurs after complete surgical excision.

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