

A 9-year-old Girl with Fibrous Hamartoma of Infancy

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ABSTRACT

Fibrous hamartoma of infancy is a rare soft tissue tumor in infants and young children with a characteristic triphasic morphology, typically occurring in the axilla and rarely in other locations. A 9 years old girl presented with complaints of swelling over right arm. On examination, a mobile and non-tender lump (size: 1.5–2 cm) was identified. Local excision was performed and tissue was sent for histopathological examination. The microscopic examination revealed fibrous hamartoma of infancy. Fibrous hamartoma of infancy is a rare benign entity with limited clinical features having an excellent prognosis, therefore a proper diagnosis is mandatory in treating it.

Keywords: Fibrous hamartoma of infancy (FHI); histopathology

CASE DESCRIPTION

A 9 years old Chinese girl presented to the dermatology department of the Second Affiliated Hospital, School of Medicine of Xi'an Jiao tong University with a 3 months history of a progressively enlarging, asymptomatic dark patch on the right arm, below the shoulder joint on the anterior surface (Figure 1). On physical examination, a lump (size: 1.5–2 cm) was identified on the right arm. The lump was soft, mobile, and non-tender. The rest of the systemic examination was normal. The lesion began as a light brown skin colored small nodule which gradually enlarged over a period of time. There is no family history of similar skin lesions. Her general physical and systemic examinations were unremarkable.

The ultrasonographic examination revealed a heterogeneous soft tissue mass along the anterior aspect of proximal right arm. The nodule was not attached to the overlying skin or underlying soft tissues, and, there was no evidence of necrosis or calcification. By considering all the clinical and ultrasonographic features, a diagnosis of FHI was concluded. The mass was therefore decided to be excised and sent for histopathological examination. Histologically, the tissue fragments disclosed an intimate admixture of fibroblastic cells and mature adipose tissues. Occasionally, some fragments showed fibroblastic cells floating in a collagenous and myxoid back ground. The dissociated cells were spindle-shaped, with scanty to moderate amount of eosinophilic cytoplasm. The histological examination confirmed the diagnosis.

Fig. 1: Right arm tumor, anteriorly below the shoulder joint.



Fig. 2: FHI composing predominantly of mature adipose tissue, fibrous trabeculae and spindle-shaped cells, in an organoid pattern (H&E ×50)

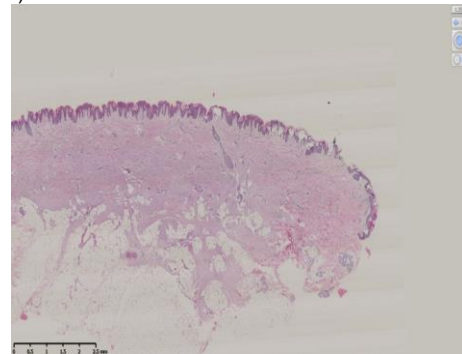
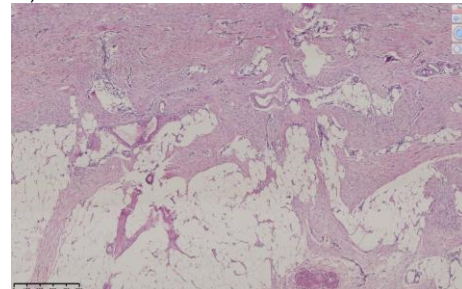


Fig. 3: FHI composing predominantly of mature adipose tissue, fibrous trabeculae and spindle-shaped cells, in an organoid pattern (H&E ×200)



DISCUSSION

Fibrous hamartoma of infancy (FHI) is a rare soft tissue tumor in infants and young children, which was first described as “subdermal fibromatous tumours of infancy” by Reye in 1956¹, and Enzinger renamed it into fibrous hamartoma of infancy in 1965². Most commonly, it occurs in children less than 2 years of age, and presents as a congenital lesion in about 20% of the cases³. The tumor is much more common in males than female, and most often present as a painless subcutaneous mass involving the axilla, trunk, upper arm, and external genitalia, although

isolated cases of fibrous hamartoma of infancy have been reported in a wide variety of anatomical locations, including the scalp⁴, foot⁵, hand⁶, and buttock⁷. MRI reports usually show an arrangement of fat with heterogeneous soft tissue bands interspersed in an organized manner, that may suggest this diagnosis in the appropriate clinical setting⁸. The differential diagnosis of fibrous hamartoma of infancy is broad, and dependent on the relative proportion of fat, mature fibrous tissue and primitive mesenchymal elements. Predominantly, fatty tumors should be distinguished from lipofibromatosis, the 'lipofibromatosis-like neural tumor'⁹ and maturing lipoblastoma. One important morphological feature of fibrous hamartoma of infancy that we believe should be given special attention to, are areas resembling another (potentially more aggressive) pediatric soft tissue tumor, such as giant cell fibroblastoma. These areas are characterized by dense, hyalinized collagen, cracking artifact, and pseudoangiomatous, slit-like spaces lined by flattened and occasionally more prominent, CD34-positive fibroblastic cells. The overall clinical features of the patients in fibrous hamartoma of infancy are similar to those previously reported, with a strong male predominance, a mean patient age of 15 months, and the involvement of the axilla and other common areas (back, upper arm, chest wall, and external genital region). We have found that the local recurrences are generally at a low rate.

Conflict of Interest: Authors have no conflict of interest.

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