Fibrous Hamartoma of Infancy: A Case Report

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Sir,
Fibrous hamartoma of infancy (FHI) is a benign but persistent soft tissue tumour that appears during the first 2 years of life. We report an unusual case of FHI from Nepal.

CASE REPORT

A 10-year-old Nepalese boy presented with a 5-year history of a progressively enlarging, asymptomatic tumour on the left side of the face. The lesion began as a skin-coloured nodule, which gradually enlarged over a period of time. There was a history of recent rapid enlargement after the incisional biopsy. No history of similar skin lesion was found in the family. His general physical and systemic examinations were unremarkable.

The cutaneous examination revealed a solitary, firm, pedunculated, non-tender, 10×8 cm^2 tumour with lobulated surface over the left parotid region (Fig. 1). The tumour was softened and ulcerated in some areas; and was associated with sero-sanguinous discharge and thick crust. The overlying temperature was normal. The tumour was not fixed to the underlying tissues. The oral examination did not reveal any abnormalities.

The differential diagnoses of pilomatricoma and parotid malignancy were made. Wedge biopsy was performed for histopathological examination but it showed only myxoid tissue; therefore, it was difficult to draw a final conclusion. Keeping in mind the differential diagnoses, the tumour was subsequently excised completely and sent for histopathological examination.

Fig. 1. Single, firm, pedunculated tumour with lobulated surface over the left parotid region.

Gross examination of the specimen revealed a poorly circumscribed tumour containing an intimate mixture of firm grey-white tissue and fat. The cut surface showed multiple small and large cystic and solid areas. Cystic spaces were filled with mucoid material.

Histopathological examination revealed a characteristic organoid pattern composed of intersecting trabeculae of fibrous tissue and collagen. There were loosely textured areas of mucoid matrix with spindle-shaped, round or stellate cells and interspersed mature adipose tissue. No muscular elements or cell atypia were observed. These features were consistent with a diagnosis of fibrous hamartoma of infancy. No recurrence was noted during the next 6 months of follow-up.

DISCUSSION

FHI is a rare benign but persistent soft tissue tumour first described by Reye in 1956 (1) as 'subdermal fibromatous tumours of infancy'. The tumour is a hamartoma because it is composed of benign disorganized proliferating tissue that occurs normally at the site in which it arises. In 1965, Enziger (2) renamed the disorder FHI. The lesion usually develops in the first 2 years of life but it can be encountered in children (3). Males are more commonly affected than females (4). The tumour occurs predominantly on the trunk, especially the axilla and upper extremities (5), but it has also been reported infrequently on the distal extremities, head, neck and scrotum (6, 7). Mostly it presents as a solitary lesion but multiple lesions have also been reported (8).

The tumour generally presents as a slowly growing, firm, painless subcutaneous mass that is usually freely movable but occasionally it is fixed to the underlying subcutaneous tissue (9). A few cases have had overlying skin changes, including alteration in pigmentation, eccrine gland hyperplasia and increased hair (8). The tumours can become as large as up to 15 cm, but most of them are 2.5–5 cm at their largest dimension (10). There is no evidence of increased familial incidence or of associated malformations or other neoplasms (5).

Although immunostaining techniques may help in differentiating FHI from malignant tumours, histopathology is pathognomic for the diagnosis of FHI (4, 9). It consists of three different elements: dense mature fibro-collagen, loose immature myxoid mesenchymal and mature adipose tissue, which forms a vague,

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irregular organoid pattern. Sometimes, the relative abundance of one kind of tissue may cause diagnostic error. Therefore, performing adequate tissue sampling is important to eliminate the diagnostic error.

Local excision of the tumour is curative and is the treatment of choice. The recurrence rate is low (4). This case has been reported for its rarity, uncommon site and the unusual size of the tumour, and late age of onset.

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