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AGGRESSIVE INFANTILE FIBROMATOSIS

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An 8-month-old infant presented with 2 ulcerated indurated plaques in the right lower limb. The lesions grew rapidly initially but for the last 2 months, they have been stationary. Biopsy showed proliferation of fusiform fibroblasts some of which were arranged in whorls. The clinical picture and the histopathology suggested the diagnosis of an aggressive infantile fibromatosis. This is one of the types of juvenile fibromatoses, a group of tumours characterised by fibrous tissue proliferation present at birth or acquired early in childhood.

Key Word : Fibromatoses

Introduction

The fibromatoses are a heterogenous group of disorders that are linked only by their biological behaviour. They represent a hyperplasia of soft tissue elements such as fibroblasts and myofibroblasts, that may be capable of local growth but are incapable of metastasizing and thus are borderline between benign and malignant. Fibromatoses may be classified into three categories: superficial (fascial) types, deep (musculoaponeurotic) types and juvenile types.[1]

Case Report

An 8-month-old infant was brought with two large masses in the right thigh and upper calf. The masses had started at the age of 2 months, grown rapidly for 4 months and were then quiescent for the past two months with no increase in the size. On examination, two large plaques were seen with a central ulceration. The larger plaque which was in the upper part of the thigh measured 15 cm x 9 cm and showed a central linear ulceration. (Fig.1). It was firm in consistency and was not tender. The smaller plaque, which was seen extending upto the upper calf, measured 8 x 5 cm and also showed central ulceration. Near both the plaques, two small erythematous papules were seen measuring about 1 x 1 cm. They were firm in consistency. A clinical diagnosis of histocytoma was made.

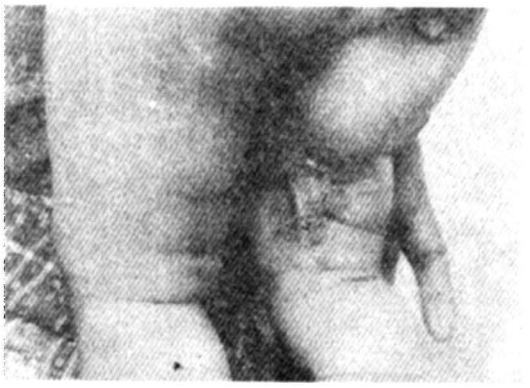

Fig. 1. Ulcerated plaque over right thigh.

Routine investigations were normal. X-ray chest was normal. X-ray of the lower limbs showed no bone involvement. On biopsy, numerous fascicles of fusiform fibroblasts were seen, some of which showed a whorled pattern (Fig 2). This localized proliferation of fibroblasts was suggestive of a juvenile fibromatosis.

Discussion

Juvenile fibromatoses is a group of conditions characterised by fibrous tissue

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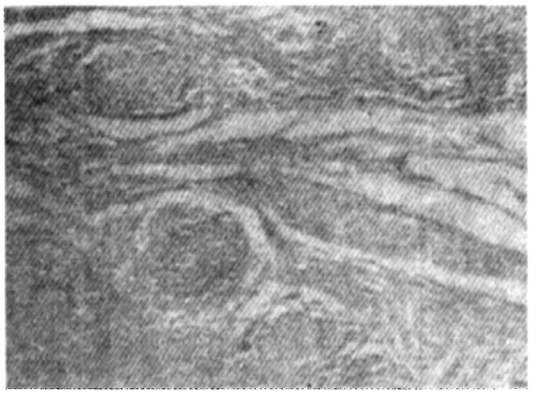

Fig. 2. Photomicrograph showing cords of fusiform cells showing a whorled pattern (H&Ex60).

proliferation either present at birth or acquired in early childhood.[2] The term juvenile fibromatoses was first suggested by Stout[3] to designate benign proliferating and infiltrating fibroblastic tissue occurring in children less than 16 years of age. These benign infiltrating growths can be divided into various subtypes.[4] They include:

1. Congenital generalised fibromatosis
2. Aponeurotic fibroma
3. Infantile digital fibromatosis
4. Diffuse infantile fibromatosis
5. Fibrous hamartoma of infancy
6. Aggressive infantile fibromatosis
7. Fibromatous colli.

Aggressive infantile fibromatosis is characterised by the presence of a single or multiple asymptomatic, firm subcutaneous swellings which show rapid increase in size. There is no predilection for this tumour which usually occurs during the first year of life. Most tumours occur on the upper and lower extremities and the trunk.[5]

Histologically, this lesion can resembe a fibrosarcoma. Fascicles of fusiform cells with varying amounts of reticular and collagen fibres are found growing in the muscle, fascia, tendons, and subcutaneous tissue. The histological changes with numerous mitosis reflect the rapid growth of the lesion. Interlacing cords of spindle-shaped cells often form a whorled pattern and reticulum fibres are prominent.[6] Clefts of carcinous spaces may be seen amidst the cells. There are also scattered round cells or mast cells. Our case showed interlacing fascicles of fusiform cells some of which showed a whorled pattern suggestive of this condition. On electron microscopy, two types of cells are identified: embryonic fibroblasts having a simple structure and more complicated cells with pinocytosis.

The course of these tumours is variable. Some are locally aggressive and need surgical excision. Others regress spontaneously. In our case, the tumour has been quiescent for the past two months and has shown no evidence of the rapid growth in the preceding 4 months.

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