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Clinical Records

Infantile fibromatosis

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Abstract
A case of infantile fibromatosis involving the cheek of a three-year-old Sudanese girl is described. The tumour was resected through a sublabial approach.

Key words: Fibromatosis, gingival; Child

Introduction

The fibromatoses comprise a group of lesions with certain common clinical and histological features (Mackenzie, 1972). These lesions infiltrate widely and replace muscle, fat and even bone with fibrous tissue of varying cellularity. The process can be diffuse, multifocal or localized in extent and infiltration spreading far in advance of the palpable margins of the growth.

Fibromatosis is an apparently autonomous proliferation of myofibroblasts, occasionally forming tumour-like masses. The myofibroblasts are locally infiltrative and may mature into dense collagenous scar-like tissue. Occasionally spontaneous 'cure' occurs (Underwood, 1992).

The commonest of this group of diseases, palmar fibromatosis causes a flexion contracture of the fingers and palm (Dupuytren's contracture). Other sites for fibromatoses include muscle (desmoid tumour), the retroperitoneum (retroperitoneal fibrosis) and the penis (Peyronie's disease).

Although some of the lesions may be precipitated by hormones or drugs, their precise aetiology and means of control are unknown.

Most fibrous tumours in childhood are benign fibromatoses (92 per cent) but some cases are malignant (Coffin and Dehner, 1992). Infantile fibromatosis represents the childhood counterpart of muscular aponeurotic fibromatosis, and arises as a solitary mass in skeletal muscles or in the adjacent fascia, aponeurosis or periosteum (Enzinger and Weiss, 1995).

It chiefly affects children from birth to eight years of age and is slightly more common in girls. There are considerable variations in the morphological appearance ranging from primitive mesenchymal forms to lesions that closely resemble adult desmoids, except perhaps for a less uniform pattern and a greater degree of cellularity.

Stout (1954) was the first to identify and describe this form of fibromatosis as a distinct entity, but since his report only a small number of cases have been added to the literature.

Here we report a case treated at our institute together with a review of the literature.

Case report

A three-year-old Sudanese girl presented to our clinic with a painless lump in the left cheek region. The parents

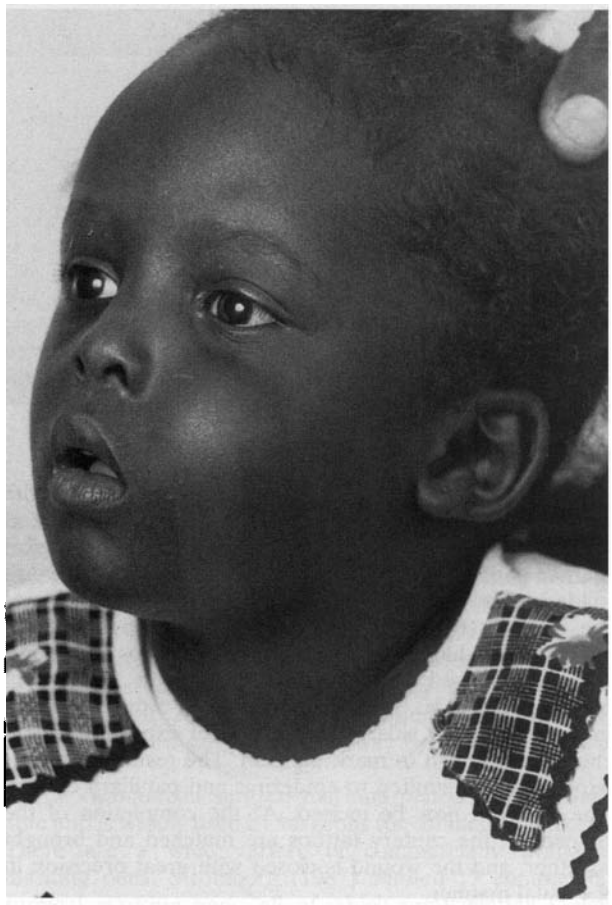

FIG. 1
Pre-operative view.

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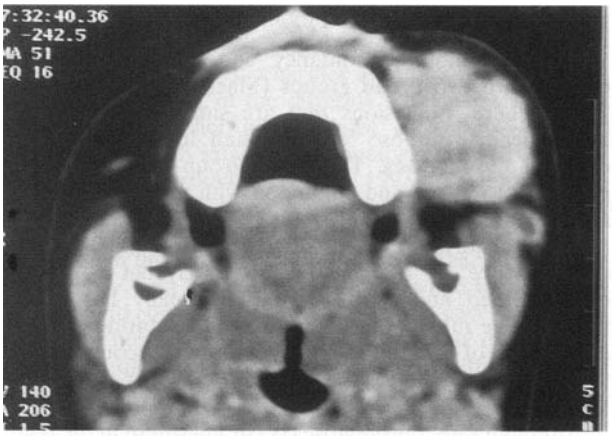

FIG. 2

Shows a well-defined homogenous soft tissue mass about 2 cm in size over the left maxilla. Following i.v. contrast there is mild diffused enhancement of the lesion.

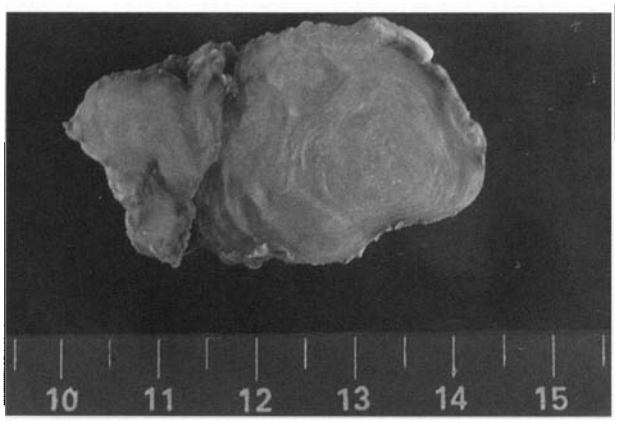

FIG. 3

Gross appearance of the specimen. Note the scar-like grey white coarsely trabeculated cut surface, the characteristic appearance of infantile (desmoid-type) fibromatosis.

had noticed the presence of the lump when she was around four months old. It had been increasing slowly and becoming more prominent. Excisional biopsy done in Sudan revealed a benign mass. Examination of the lump revealed a firm, nodular lump (4 × 5 cm), with a slightly irregular surface (Figure 1). The lump was non-tender. The facial nerve and the trigeminal nerve were intact pre-operatively. Head and neck and systemic examination were normal. Computerized tomography (CT) scan showed a soft tissue mass in the left infraorbital region with no calcifications and no cystic or fat components (Figure 2).

The lump was excised through a sublabial incision. Local injection (lidocaine 36 mg + adrenaline 0.0225 mg) was given on the area of incision (4–5 cm overlying the mass). Due to the size of the lump bleeding was anticipated pre-operatively, and thus consent for possible intra-operative or post-operative blood transfusion was taken. Intra-operatively, the mass was found not to be capsulated, with poor demarcation. Involvement of the surrounding soft tissues of the cheek could not be ruled out, but there was no maxillary bone involvement. The intra-operative bleeding was minimal, and

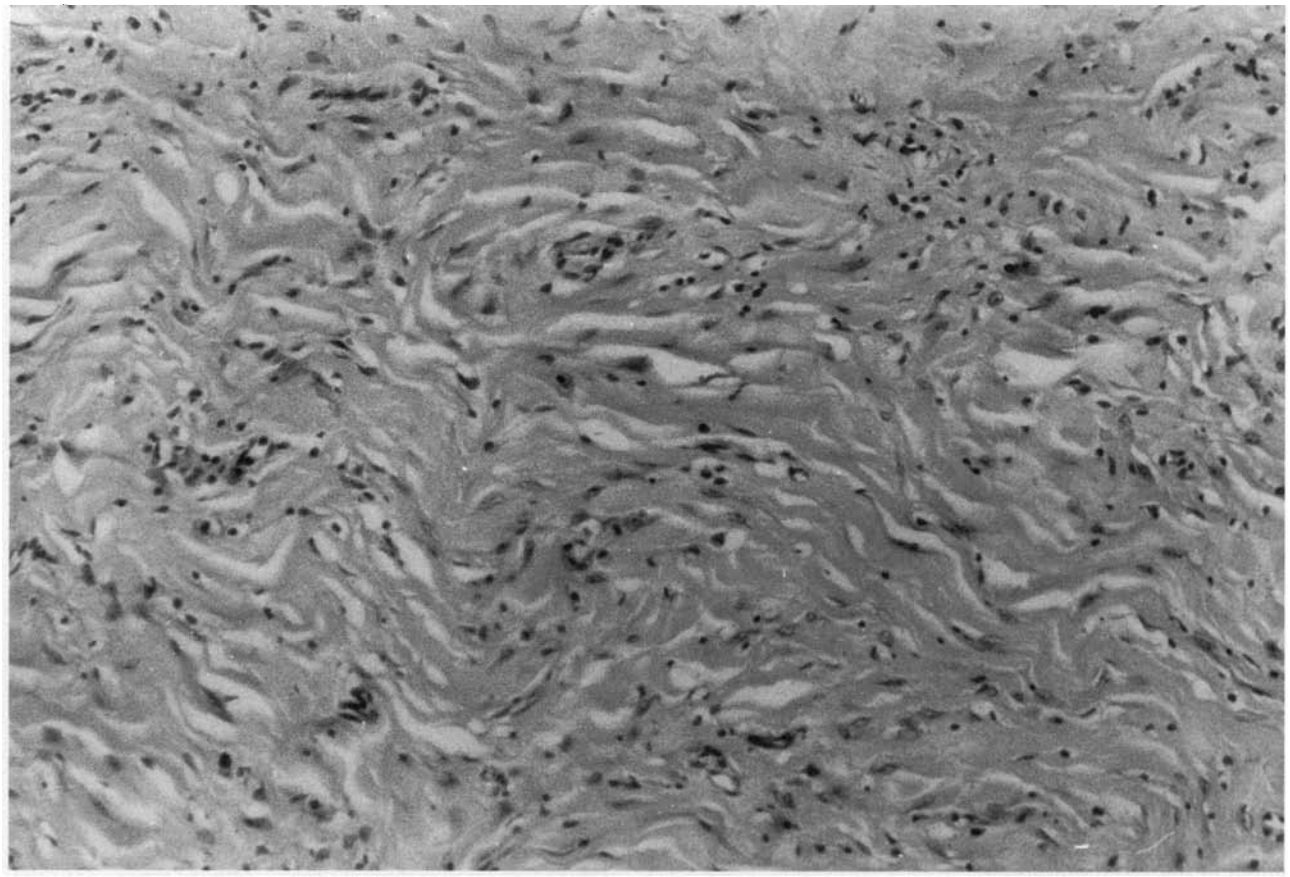

FIG. 4

Microscopic appearance of infantile fibromatosis showing proliferation of fibroblasts with deposition of variable amount of collagenous tissue and mononuclear cell infiltrate (H & E; × 200).

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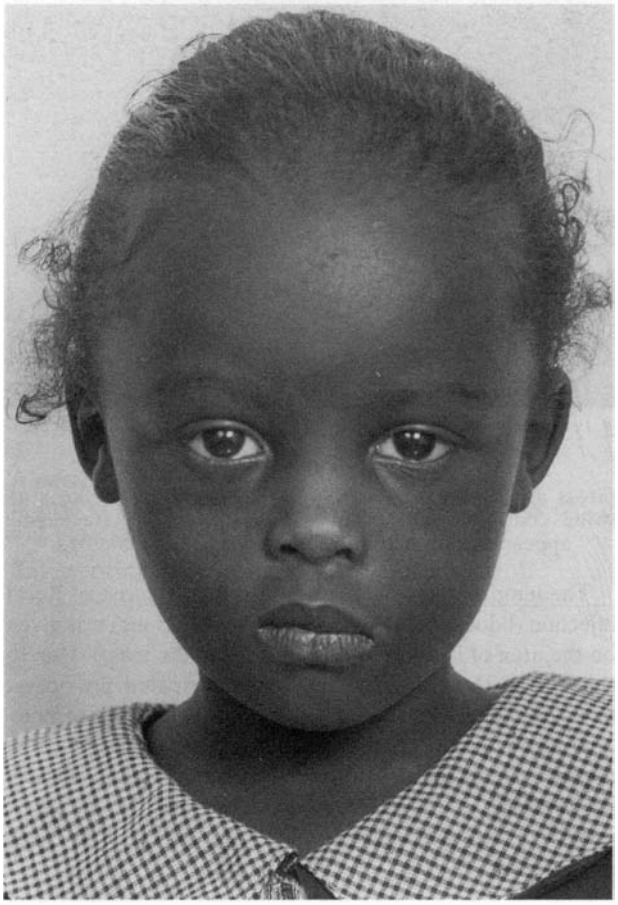

FIG. 5
Post-operative view, with mild residual left-sided facial weakness.

thus blood transfusion was not required. The defect was closed using absorbable sutures. No monitoring of the facial nerve was carried out intra-operatively.

Macroscopically it was a firm nodular lump with extensive adhesions to the surrounding connecting tissue (Figure 3). Microscopic examination showed fibroblastic proliferation, surrounded by thick collagenous tissue (Figure 4), with neither bone elements or calcification noted. The findings were consistent with fibromatosis.

Post-operatively, the girl developed swelling of the cheek which subsided within 10 days. She was also found to have a residual left-sided facial weakness involving the buccal branch which was manifested by limitation of movement of the upper lip. The weakness was most probably provoked by the excessive stretching intra-operatively. The weakness improved with time and was back to normal within two months (Figure 5).

Also as a consequence of the excessive stretching intra-operatively, during the immediate post-operative period numbness sensation was noted on the involved cheek and lateral side of nose as well as the upper lip. This sensation disappeared within a few days, with no residual defects noted in the facial sensations.

The child is followed up regularly for the possible chance of recurrence of the mass due to the poor demarcation noted.

Discussion

Fibrous tumours of infancy and childhood can be divided into two broad groups (Mackenzie, 1970; Allen, 1977). There are lesions that are similar clinically and histologically to those lesions found in adults such as desmoid fibromatosis. The other group includes lesions peculiar to infancy and childhood with no counterpart in adult pathology.

In the majority of cases, treatment is sought because of the presence of a firm solitary mass that is poorly circumscribed and deep-seated and usually has grown rapidly. The mass is nearly always noted during the first eight years of life and is most commonly encountered in the first and second year after birth (Enzinger and Weiss, 1995).

Although rare, fibromatoses occur anywhere in the body and maybe superficially or deeply placed (Carr *et al.*, 1992). In most cases the mass originates in the skeletal muscle, especially in the muscles of the head and neck (34 per cent), the shoulders and upper arm (32 per cent), the trunk (18 per cent) and the thigh (16 per cent) (Enzinger and Weiss, 1995).

In the head and neck region, the preferred sites are the tongue, the mandible, the maxilla, and the mastoid process (Hidayat and Font, 1980; Shah and Katz, 1988; Tagawa *et al.*, 1989; Thompson *et al.*, 1991). Primary involvement of the oral cavity, nasal cavity and paranasal sinuses is much less common, which is fortunate as these lesions seem to be more destructive and lethal than at other sites (Fu and Perzin, 1976). As the lesion progresses, it may infiltrate adjacent muscles and may grow around vessels and nerves, resulting in tenderness, pain or functional disturbances.

The growth rate may decrease after initial rapid growth, and periods of activity may alternate with more static periods or even arrested growth. They may even regress spontaneously (Allen, 1977). As many as 50 per cent of cases grow progressively and 50 per cent show phases of arrested growth, albeit of short duration (Carr *et al.*, 1992). They have a marked capacity to recur, even after seemingly adequate excision and this poses a major problem in eradication. Recurrence is more frequent in younger patients.

Fibromatoses are twice as common in children than adults, and 25 per cent occur in children less than 15 years of age (Conley *et al.*, 1966). Fibromatoses are more common in females than males at a ratio of 3:2 (Carr *et al.*, 1992).

Radiological examination shows a soft tissue mass sometimes associated with bowing or bone deformation. In fact, sometimes it is difficult to detect whether the lesion arose in the soft tissues, periosteum or bone (Melrose and Abrams, 1980; Rodu *et al.*, 1981), thus included in its differential diagnosis are cases of ossifying fibromas and fibrous dysplasia. The latter diseases are fibro-osseous lesions while fibromatosis is a soft tissue lesion. Histological examination along with radiological and clinical findings are very helpful in differentiating these lesions.

Grossly the lesion of fibromatosis consists of firm, ill defined, scar-like masses of gray-white tissue measuring one to 10 cm or more, in greatest diameter. It is never encapsulated and usually is excised together with portions of the involved muscle or subcutaneous fat.

Microscopically, infantile fibromatosis has a wide morphological range reflecting progressive stages in the differentiation of fibroblasts. The least mature and the most common type of lesion is often described as the diffuse or mesenchymal type. Other types are the fibroblastic type, the desmoid type and the aggressive type, which is very cellular and difficult to distinguish from infantile fibrosarcoma.

The diffuse type of this tumour can be confused with other myxoid and lipomatous tumours because of the large amount of mucopolysaccharides in the stroma and the partial replacement of the diffusely infiltrated muscles by lipocytes. The other lesions most commonly confused histologically with infantile fibromatosis are myofibromatosis and fibrosarcoma (Carr *et al.*, 1992). It is important to distinguish between these conditions as they may display quite different clinical behaviour.

Although the tumour does not metastasize, it may reach a large size, and tend to recur locally when inadequately excised, and continues to grow in a progressive, infiltrative manner. Thus, complete excision with ample margins, the treatment of choice, is often extremely difficult and in some cases may be impossible without disfigurement or dysfunction.

The locally aggressive behaviour, the infiltrative growth beyond palpable margins and high recurrence rate have prompted many to advocate aggressive surgery to eradicate these growths (Seel *et al.*, 1964). Wide 'en bloc' removal in three dimensions can be a formidable operation, especially in the child and, for neck lesions, is more easily managed by radical neck dissection (Masson and Soule, 1966).

Infantile lesions seem to be more aggressive and head and neck lesions particularly so, but this may reflect the anatomical difficulties encountered when attempting to eradicate them from this site. Head and neck lesions infiltrate more rapidly and widely (Wilkins *et al.*, 1975).

Aggressive surgery can reduce recurrence rates to 33 per cent (Hunt *et al.*, 1960), but a recurrence rate for extra-abdominal desmoids of 25 per cent with radical procedures has been reported (Das Gopta *et al.*, 1969). Thus for so-called benign disease, radical surgery would seem unacceptably mutilating and for neck disease may still fail to eradicate the lesion.

Spontaneous regression of desmoids and extra-abdominal desmoids has been reported after biopsy and incomplete removal which is coincident with both menarche and menopause (Strode, 1954; Bullock, 1955; Caldwell, 1976). These tumours also seem to cease growth after multiple recurrences if left for long enough (Enzinger and Shiraki, 1967).

The cause of infantile fibromatosis is not yet clear. So far there is no strong evidence that trauma or even familial predisposition play any significant role in its pathogenesis (Enzinger and Weiss, 1995).

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