

JUVENILE APONEUROTIC FIBROMA

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SUMMARY: Juvenile aponeurotic fibroma is a rare benign fibroblastic tumour usually found in young children, with a tendency to develop in the palms and soles and a high recurrence rate without any metastatic spreading. About 77 cases have been reported in the literature by 1988. A new case with recurrence is presented and the literature reviewed.

Juvenile aponeurotic fibroma or calcifying fibroma, first described by Keasbey in 1953, is a form of juvenile fibromatosis (1). It is an uncommon benign fibroblastic tumour usually found in young children. It has a tendency to develop in the palms and soles, definite histologic features of an ill-circumscribed fibroblastic proliferation with calcifications and cartilage differentiation and a high recurrence rate without any metastatic spreading. About 77 cases of juvenile aponeurotic fibroma have been reported in the literature by 1988 (2,3,4,5).

CASE REPORT

A 3.5 year old boy presented to our clinical with a gross mass in the palm of his right hand. The mass, about the size of a small orange (7 cm. in diameter) was firm, mobile, painless and enlarging slowly (Fig. 1). X-ray films showed no bony involvement. Routine blood examinations and bone marrow aspiration were of no significance. It was removed by conservative surgical excision followed by an uneventful recovery and the microscopic examination revealed a diagnosis of aponeurotic fibroma (Fig 2,3). After a period of missed clinical appointments, 8 months later, the patient admitted to our clinic with a mass of 3 cm. in diameter in his same palm (Fig 4). Again by conservative surgical excision the mass was removed and the microscopic diagnosis was the same: aponeurotic fibroma. Up to date close follow-up

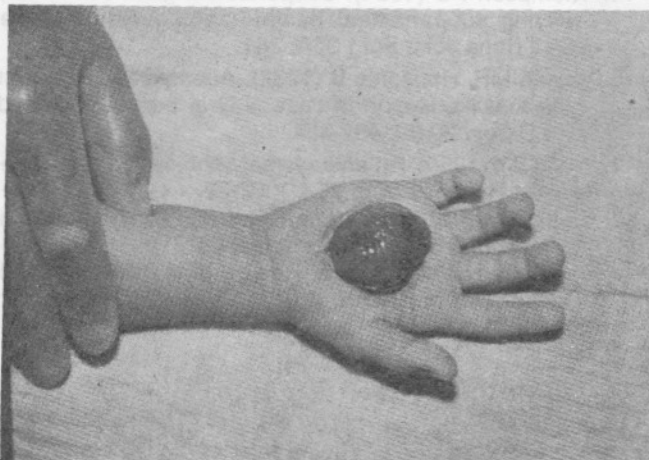


Figure 1: Per-operative view of the patient.

of the patient revealed no recurrence.

DISCUSSION

Juvenile fibromatoses are characterised by Stout in 1954 (6). Among the several clinico-pathologic forms, the first is the juvenile aponeurotic fibroma (JAF) described by Keasbey in 1953 (2,3,4,5). It is a distinctive fibroblastic tumour with three basic characteristics: a tendency to devel-

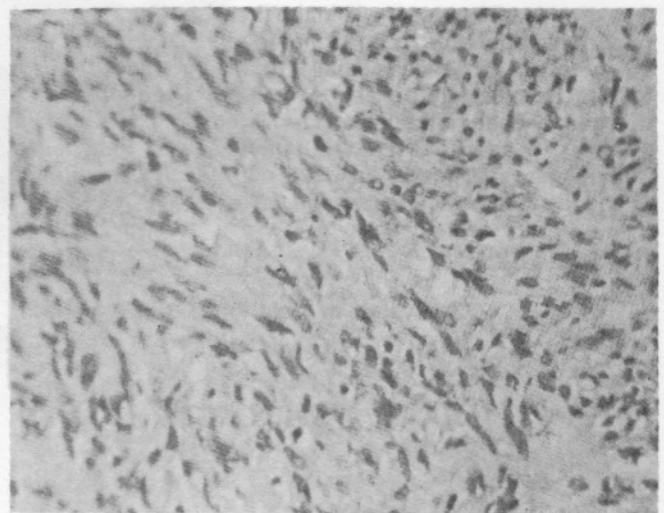
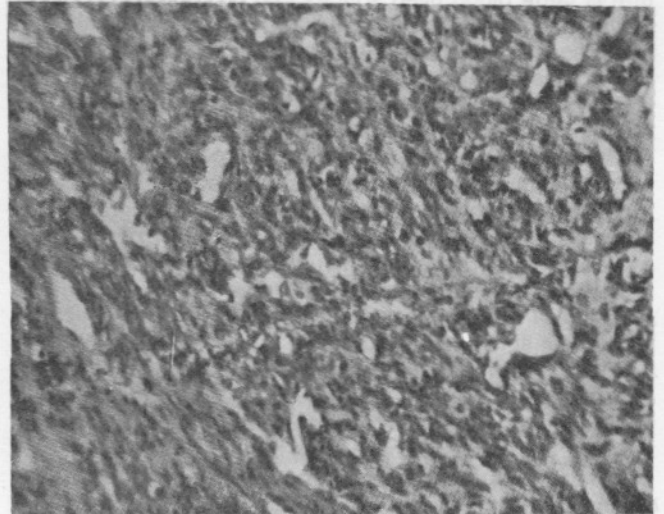


Figure 2,3: Photographs of the microscopic sections of the specimen.



Figure 4: The same patient 8 months later, presenting recurrence.

op in the extremities of children and young adults, most frequently in the palms and soles although it has been reported to occur congenitally (7), as well as in adults over 30 years of age (8,9,10), the definite histologic features of an ill-circumscribed fibroblastic proliferation with calcification and cartilage differentiation, as it was in our case and a high recurring pattern without metastatic spreading. It is a locally invasive tumour and has a high tendency to recur but no malignant degeneration has been reported (2,3,4,5) except a case report of an aponeurotic fibroma with a fatal pulmonary fibrosarcoma by Eisenbaum (5). Recurrence has been seen as early as 6 months or as late as 6 years (5). It has a selflimiting potential which increases with age (2,4,5).

In the hand JAF is almost invariably located in the palm. Only two cases in the literature were reported to appear on the dorsum of the hand (2). Histologically JAF has been considered a cartilage analog of fibromatosis (4,5). Liechstein and Goldman (8,9) have first emphasized the cartilagenous components of the tumour. They believed that although the fibroblastic proliferation was significant, the

distinguishing and unique feature of the lesion was the appearance of focal chondroid differentiation within a spindle cell proliferation. The lesion therefore appears to represent a type of fibromatosis comprising specialised mesenchymal elements capable of differentiation towards fibrocartilage, Iwasaki et al. (10) have verified the cartilagenous nature of the tumour by ultrastructural studies with EM? Briefly it can be stated that JAF probably originates from fibroblasts that have a capacity to differentiate into cartilage and fibrous tissue.

JAF can follow a benign or an aggressive course. It must be differentiated from fibrosarcoma to avoid unnecessary radical surgery. Generally a wide excision of the tumour without sacrificing and functionally important structures will probably give the greatest chance for cure. In the event of recurrence further local wide excisions without sacrifice of any functional tissue is still indicated and a careful follow-up is mandatory.

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...Leyonun mikroskopik incelemesinde ...
 ...mevzii artıktakı zıtıye uđı ...
 ...tu uzanan proliferatı fibroblast ve ...
 ...dokunu kılantın (Fazın 1) ve ...
 ...çıkıı, amfolitik slobokzman gang ...
 ...Fazın 2) ...
 ...dev hücreler izlendi (Fazın 3) ...
 ...kılavı ve orta derecede pleomorf ...
 ...da pleomorfizm ve sellülerite ...
 ...gıoblastik mızozite tanısı konuldu (Fazın 4)

TARTIŞMA

...ilk olarak 1953 yılında yayınlandı. 7 ...
 ...Kern (8) tarafından tanımlanan proliferatlı mızozite çizgili ...
 ...Kern tarafından ilk olarak görülen pleomorfizm ...
 ...sıradır. Çok hızlı büyüme ve metastatik ...
 ...ile birçok yavaş yavaş büyüyen ...