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Aggressive Fibromatosis*

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Abstract. Four patients with aggressive fibromatosis are presented as illustrative examples of this rare non-metastasizing fibroblastic soft tissue tumor. The skeleton was involved in all four of these cases. The radiographic and histological appearances are reviewed and a summary of the literature is presented.

Key words: Aggressive fibromatosis – Soft tissue tumors – Fibroblastic tumors, fibromatoses

The fibromatoses are a group of non-metastasizing fibroblastic tumors which tend to invade locally and recur after attempted surgical excision [1]. The group includes such essentially benign conditions as Dupuytren's contracture and Peyronie's disease, as well as such aggressive conditions as desmoid tumors and so-called "congenital fibrosarcoma" (Table 1). Agressive fibromatosis is a term which is now used to describe that group of tumors which had previously been classified as musculo-aponeurotic fibromatosis, extra-abdominal desmoids [2], and periosteal desmoids [6]. It is also probably appropriately applied to some cases of desmoplastic fibroma involving the skeleton [7]. The authors have had experience with 10 proven cases of aggressive fibromatosis in Rochester over the past 10 years. The radiologic changes which may be associated with these lesions have seldom been recorded. In this report four cases demonstrating radiologic evidence of bone involvement are discussed and the radiologic and pathologic features are illustrated.

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Table 1. The fibromatoses

"Adult" fibromatoses
1. Dupuytren-type fibromatoses
   a) Palmar fibromatosis
   b) Plantar fibromatosis
   c) Peyronie's disease
2. Desmoid fibromatoses
   a) Extra-abdominal desmoids
   b) Abdominal wall desmoids
   c) Intra-abdominal desmoids
   d) Multiple desmoids
   e) Gardner's syndrome

"Juvenile" fibromatoses
1. Congenital fibrosarcoma-like fibromatosis
2. Congenital localized or generalized fibromatosis
3. Juvenile aponeurotic fibroma
4. Fibrous hamartoma of infancy
5. Recurring digital fibrous tumor of childhood

Case Reports

Case 1

This 17-year-old boy (B.D.) presented at an outside hospital with a mass in his antecubital fossa which was excised. Six months later in July 1978, there was a recurrence at the wrist on the same arm and he was referred to the Strong Memorial Hospital. He presented with a large soft tissue mass just above the wrist which was not attached to the skin. Radiographs at the time were read as normal but in retrospect there may have been some periosteal erosion (Fig. 1). The mass was excised with an apparent margin of normal tissue. He reappeared in December 1978 with a recurrence of the mass spreading up almost to the elbow. Radiographs taken at this time revealed widespread bony involvement of the radius and ulna (Fig. 2). He had his arm disarticulated above the elbow. In May 1979, he reappeared with a recurrence in the upper arm and shoulder girdle (Fig. 3), and disarticulation of the arm at the shoulder was performed. Unfortunately at operation, involvement of the muscles of the thorax and shoulder girdle was found, however he has had no recurrence to date.

Case 2

In June 1978 this 58-year-old female (M.C.) fell on her outstretched hand and sustained a fracture of the distal radius

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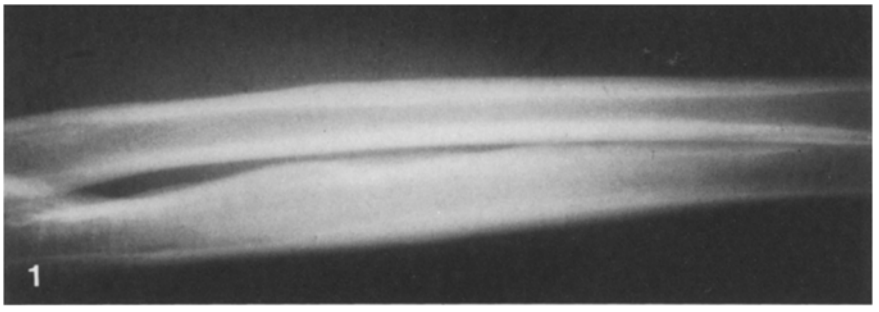
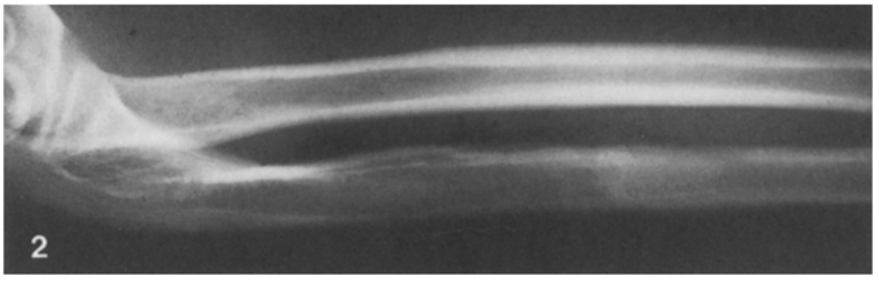




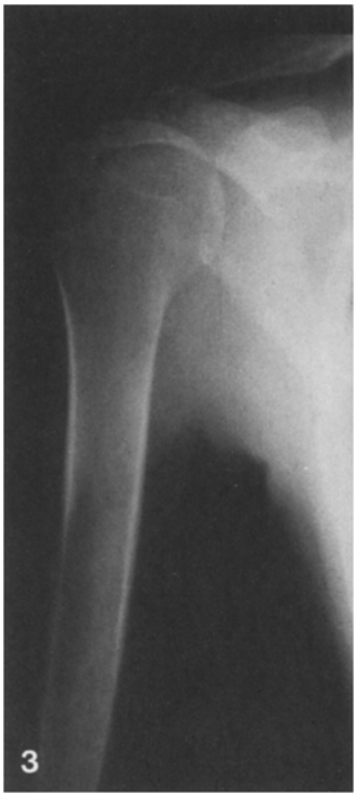


Fig. 1. *Case 1.* July 1978. Well organized periosteal new bone formation is present on the ulna aspect of the radius

Fig. 2. *Case 1.* December 1978. Invasion of the radius with cortical destruction and cancellous involvement is now noted, as well as the periosteal reaction

Fig. 3. *Case 1.* May 1979. Generalized patchy loss of bone can be seen in the upper humerus which although possibly caused by disuse was found at operation to be due to direct invasion of the bone by the tumor

(Fig. 4). It was noted that this was in fact a pathologic fracture through an area of fibrous dyplasia. The fracture healed satisfactorily but she reappeared in September 1979 with a mass in the same wrist. Radiographs taken at that time showed erosion of both the distal radius and ulna (Fig. 5). She was observed until July 1980 when the erosions had become more obvious (Fig. 6). A biopsy was taken and the diagnosis of aggressive fibromatosis was made. As much of the mass as possible was removed including part of the radius and the distal ulna. In April 1981, the mass recurred (Fig. 7) and once the extent of the tumor was realized, the patient was advised to have an amputation, which she refused. The mass was again removed along with the distal radius and ulna which were replaced by a fibular graft. In February 1982 a recurrence of the tumor occurred without any bone involvement and on this occasion the patient underwent amputation of the arm.

Case 3

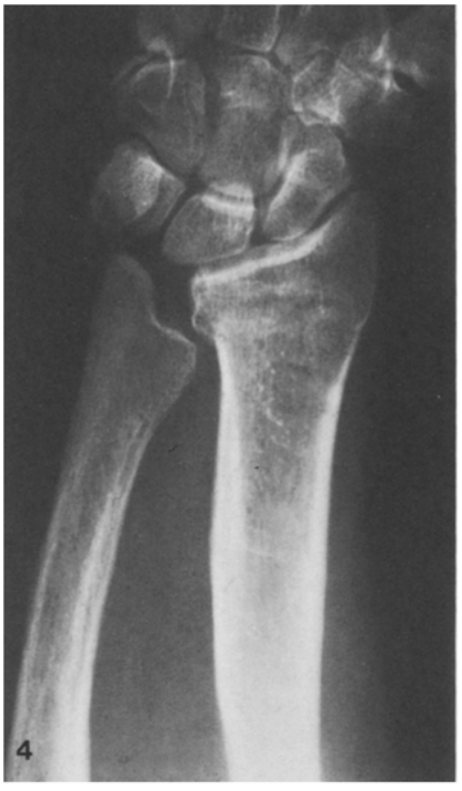
In January 1969 this 26-year-old woman (J.J.) sustained an injury to her right forearm in a motor vehicle accident. A $1^1/_2$ cm mass over the ulna was subsequently noted in May of 1969. In November of the same year, the mass which had increased in size was excised and the diagnosis of hyperplastic cicatrial fibrosis was made. The mass recurred in March 1971 with obvious osseous involvement (Fig. 8). The distal ulna and part of the radius was excised. The mass recurred in February 1972 (Fig. 9) and radiation therapy of 4,500 rads was administered. Since that treatment to the present time there has been no recurrence of the tumor and the patient has maintained surprisingly good function of the arm.

Case 4

This 94-year-old lady (R.H.) presented with a mass in her left forearm in June 1980 (Fig. 10). This was removed but the mass recurred in September 1981 (Fig. 11) when she refused operation and was referred for radiotherapy.

Discussion

The group of fibrous tissue tumors now best categorized by the term "fibromatoses" rarely have significant radiologic changes apart from soft tissue swelling or a soft tissue mass. However, the condition now known as "aggressive fibromatosis" can often locally invade the adjacent bones as our four illustrative cases show. The 10 documented patients we have seen range in age from 5 to 94 years and six are female. The arm is involved in five, the mandible, maxilla, chest wall, neck, and leg in one case each. The recurrence rate is 50% and our mortality rate is 10% due to direct local invasion of the chest wall. Three cases were treated by radiotherapy, two apparently successfully having no recurrence at the time of writing, although the follow-up period is short in one patient. Three patients were treated by radical excision and the other seven by local excision. There have been two recurrences so far in the first group



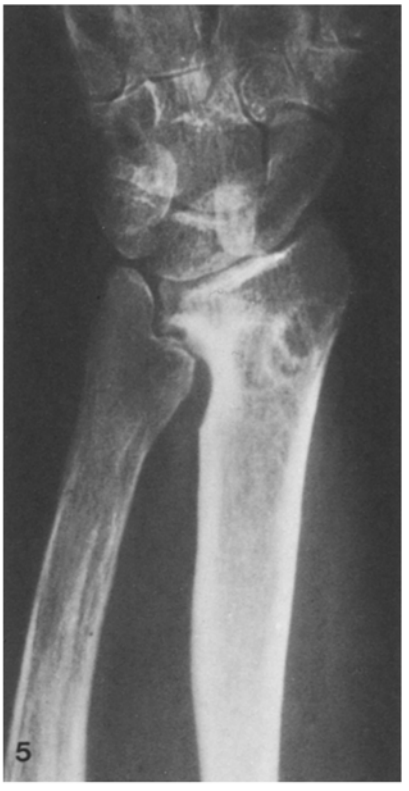
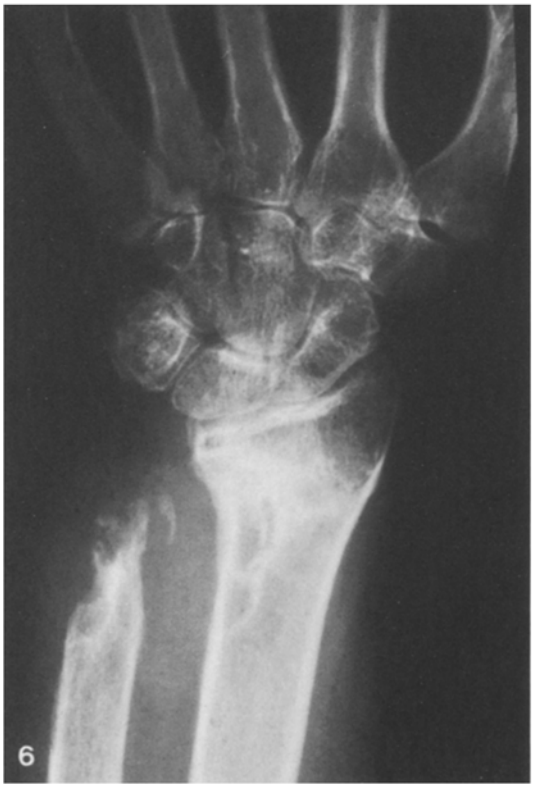


Fig. 4. *Case 2.* June 1978. The ulna is bowed and expanded with a double cortical contour and a ground glass appearance. There is a Madelung deformity. The distal radius is also symmetrically expanded and has the characteristic appearance of fibrous dysplasia. There is a fracture of the distal radius

Fig. 5. *Case 2.* September 1979. Some well circumscribed lucencies have appeared in the distal radius and erosion of the ulna aspect of the bone is noted. Disuse osteoporosis is present



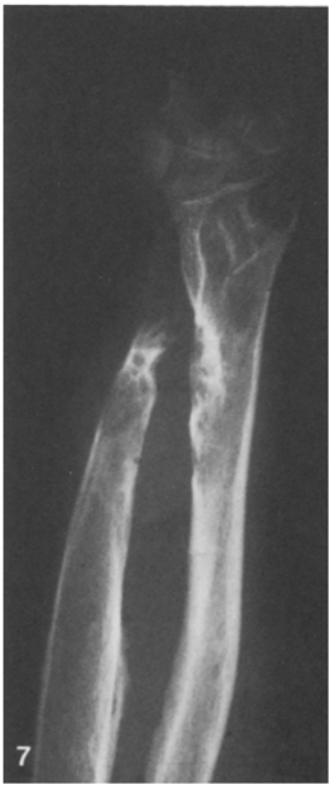


Fig. 6. *Case 2.* June 1980. Although no operation was performed, the distal ulna has now been eroded away leaving defects with sclerotic margins in both the remains of the ulna and in the distal radius. Note that some of the lucencies previously seen in the radius have apparently disappeared

Fig. 7. *Case 2.* April 1981. The tumor has become more extensive and invaded much of the distal radius as well as continuing to destroy the distal ulna. A soft tissue mass is apparent

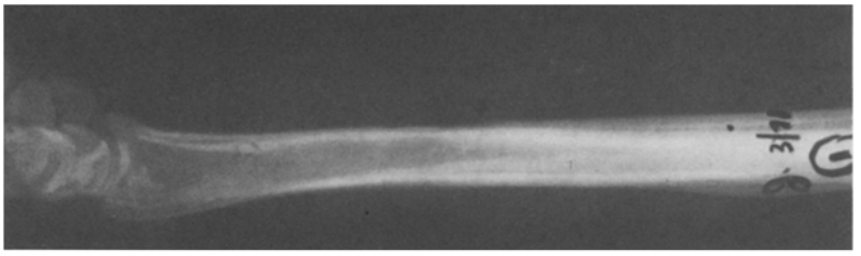


Fig. 8. *Case 3.* March 1971. A large soft tissue mass is apparent in the forearm with a well organized periosteal reaction

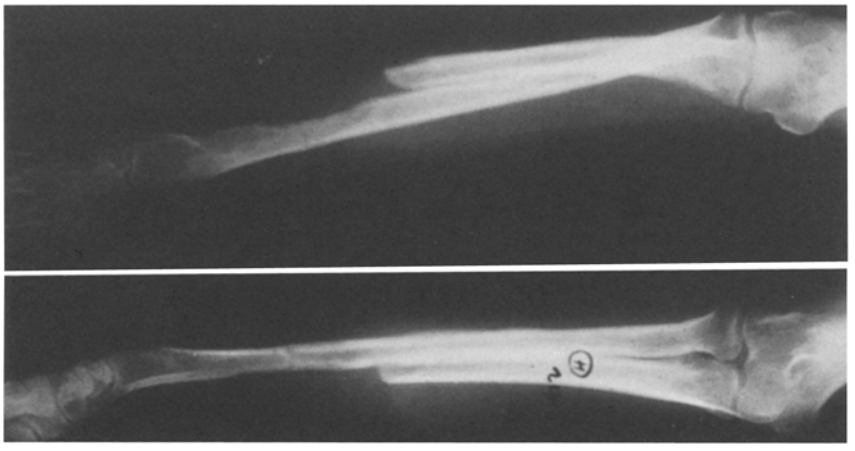


Fig. 9. *Case 3.* April 1972. Following excision of the distal ulna and the ulna aspect of the distal radius, the mass recurred with further erosion of the radius

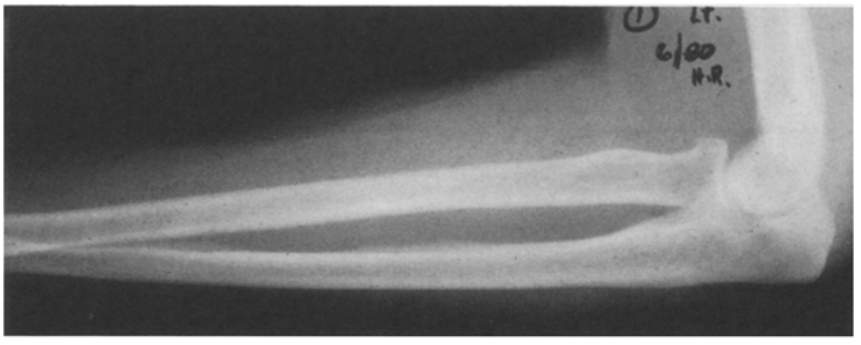


Fig. 10. *Case 4.* June 1980. A soft tissue mass over the proximal ulna is apparent without bony involvement

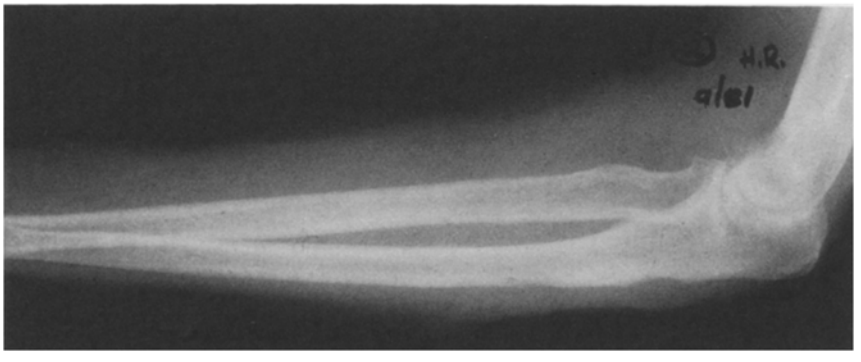


Fig. 11. *Case 4.* September 1981. External erosive changes are now apparent in the ulna as well as enlargement of the soft tissue mass

and four recurrences following local excision. On the whole these findings are consistent with those discussed in Allen's series of 140 cases which are based on his own patients [1] as well as those described by Enzinger and Shiraka [4], Das Gupta et al. [3], and Hunt et al. [5] (Table 2). It is of interest, however, that 50% of our cases occurred in the arm.

Aggressive fibromatosis is characterized by a deep seated mass which rarely involves the skin.

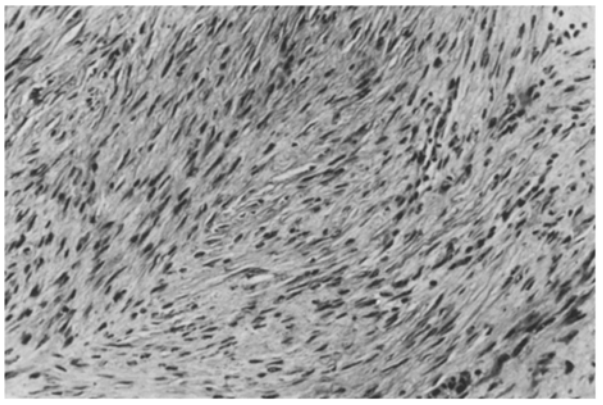


Fig. 12. Low power view (70×) demonstrating the typical appearance of fibromatosis with spindle shaped fibroblasts arranged in bundles

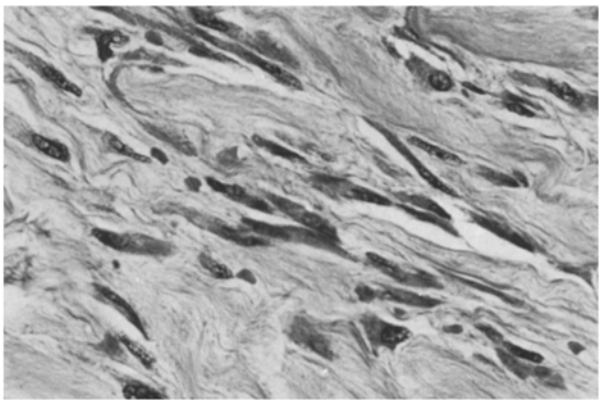


Fig. 14. In this high power view (140×) note that the nuclei are oval and elongated. No mitoses are apparent

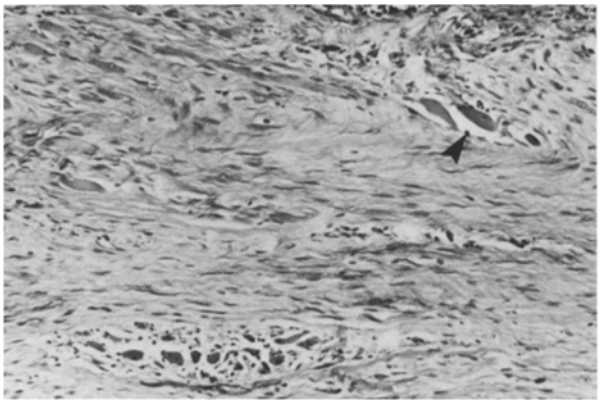


Fig. 13. Low power view (70×) showing invasion of skeletal muscle. Note residual muscle bundles (arrowhead)

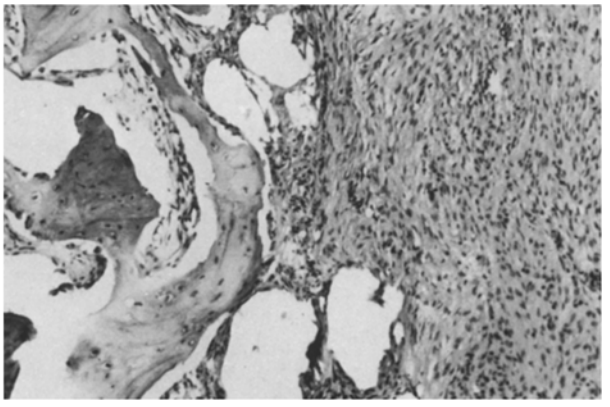


Fig. 15. In the low power view (70×) there is evidence of the tumor infiltrating and invading the bone. Note some residual trabeculae on the left of the illustration

Table 2. Characteristics of aggressive fibromatosis

Sex	Female more than male
Age	1–80 years, majority 20–30 years
Site	Arm 25–40%; trunk 10–15%; head and neck 20–30%; leg 20–30%
Recurrence rate 20–40%	
Mortality ±10%, from direct invasion of neck or chest wall	
Differential diagnosis: benign soft tissue tumors; fibrosarcoma; rhabdomyosarcoma	

Based on Allen [1], Das Gupta et al. [3], Hunt et al. [5]

Histologically the lesion consists of a moderately cellular proliferation of spindle-shaped fibroblasts arranged as interlacing bundles of cells (Fig. 12). These cells typically invade and extend between fascicles of skeletal muscle (Fig. 13). The cytologic features are generally bland. Pleomorphism is lacking and fibroblastic nuclei are oval to elongated and are not hyperchromatic: mitotic figures are rare (Fig. 14). The cellularity of the process may vary from area to area and frequently foci of dense, hyalinized paucicellular collagen may be present. Scattered inflammatory cells and focal mucoid areas may also be noted. The lack of pleomorphism and the low mitotic rate serve to distinguish aggressive fibromatosis from fibrosarcoma. Invasion of the skeleton has rarely been described although it was seen in all four of our patients (Fig. 15).

The radiologic appearance is frequently of a soft tissue tumor slowly invading the local osseous structures producing periosteal irregularity and erosion. As the tumor progresses, it may invade and destroy the whole bone. The radiologic differential diagnosis includes any soft tissue tumor invading the bone, but the lack of osseous reaction should suggest a benign lesion rather than a fibrosarcoma for example. A radiologic differential diagnosis would include many benign or nonaggressive lesions such as villonodular synovitis (Fig. 5) and malignant or aggressive tumors including

sarcoma. A biopsy will provide the correct diagnosis. One of our cases is of particular interest (Case 2) because of the association between fibrous dysplasia and aggressive fibromatosis which has not been reported previously: presumably both lesions arise from the same abnormal cell chain.

Various forms of therapy have been advocated including excisional biopsy [1, 4], radical surgery including amputation [3, 5], radiotherapy [1], and hormonal therapy particularly testosterone [5]. Spontaneous regression has been reported in a number of patients [1, 3–5]. Although aggressive fibromatosis is not a truly malignant tumor, in view of the high recurrence rate, it would seen that radical surgery is the treatment of choice once the initial diagnosis has been confirmed. If the tumor recurs then radical surgery with amputation and the use of radiotherapy appears to be the most reasonable method of controlling aggressive fibromatosis.

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