

# Desmoplastic fibroma of bone

## Case report and literature review

A case is described of the rare, benign intraosseous desmoplastic fibroma, occurring in the humerus of a 20-year-old woman. A total of 121 cases is briefly reviewed and a table of 78 references dealing with desmoplastic fibroma is given.

Treatment with indomethacin and ascorbic acid is proposed for cases of desmoplastic fibroma of bone when mutilating operations are the alternative or in inoperable patients.

*Key words:* desmoid; desmoplastic fibroma; fibroma; treatment; tumour.

In 1958, Jaffe introduced the term "desmoplastic fibroma of bone" to describe a densely fibrous benign lesion poor in cells, which had not earlier been discussed as an entity and which resembled the familiar desmoid tumour of the abdominal wall. In 1979, Lagacé et al. showed, by means of electron microscopy, that the lesion is principally characterized by myofibroblasts admixed with fibroblasts and primitive mesenchymal cells, and this was confirmed in 1981 by Chomette et al., who therefore suggested that the lesion is of non-neoplastic nature, signifying a pathologic scar.

Although Nilsson & Göthlin in 1969 by re-evaluation of all fibrous lesions admitted to Karolinska Institutet in Stockholm found as many as nine cases of desmoplastic fibroma of bone, a total of only 50 cases (reviewed by Sugiura in 1976) is generally referred to as the known number of cases. This study reviews 121 cases published since 1958 (Tables 1 and 2).

## Case report

In January 1973, a 20-year-old woman, complained of intermittent pain in the distal part of the right upper arm. During the following year the pain increased and the arm became swollen. In July 1974, a radiograph demonstrated a multicameral cystic process in the distal part of the right humerus (Figure 1). The patient was then admitted to the tumour centre. Supplementary radiographs of the skeleton and chest and laboratory studies were normal. The tumour was removed by curettage, and an autologous bone

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graft was inserted into the cavity. Microscopy showed connective tissue consisting of abundant collagen fibres and relatively few, elongated fibroblasts with uniform nuclei and without any pleomorphism or mitotic activity. After the operation, there was limitation of extension by 20 degrees of the right elbow and slight inter-



Figure 1. Radiograph taken in July 1974 showing the typical appearance of a medullary expanded multicameral radiolucency with thinned overlying cortex, resembling lesions such as giant cell tumour, medullary fibrosarcoma, osteolytic osteosarcoma or a metastatic carcinoma.

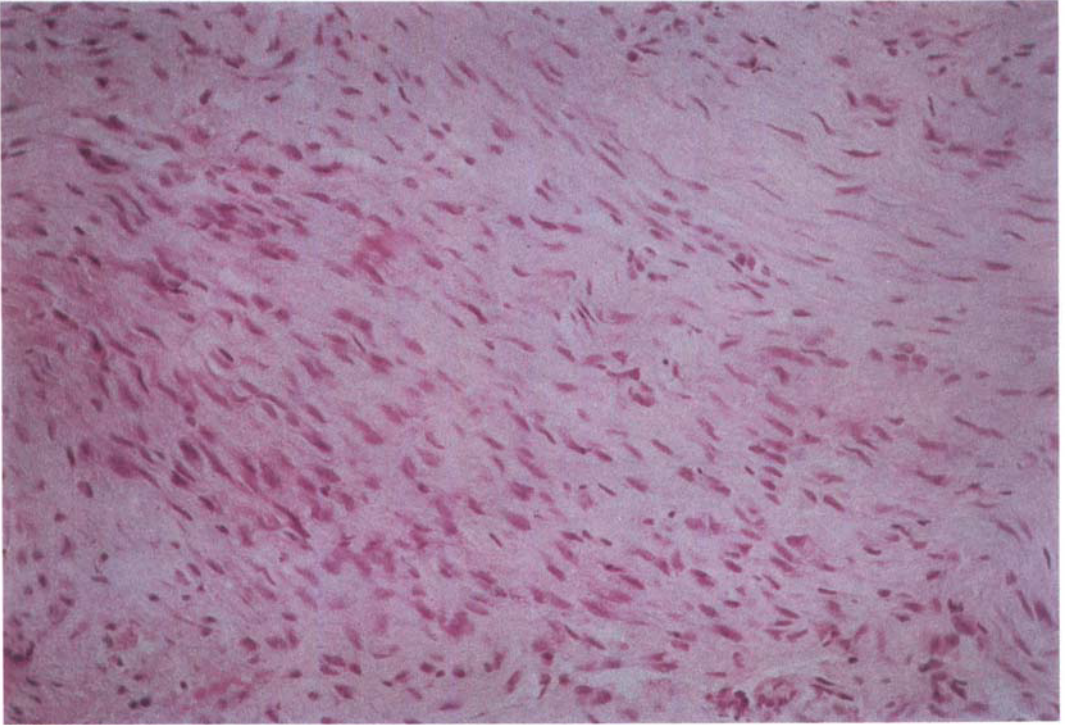


Figure 2a. Histologic specimen characterized by abundant collagen fibres and relatively few uniform cells. The cellularity, pleomorphism and mitotic activity that are features of fibrosarcoma are lacking (H and E  $\times 170$ ).

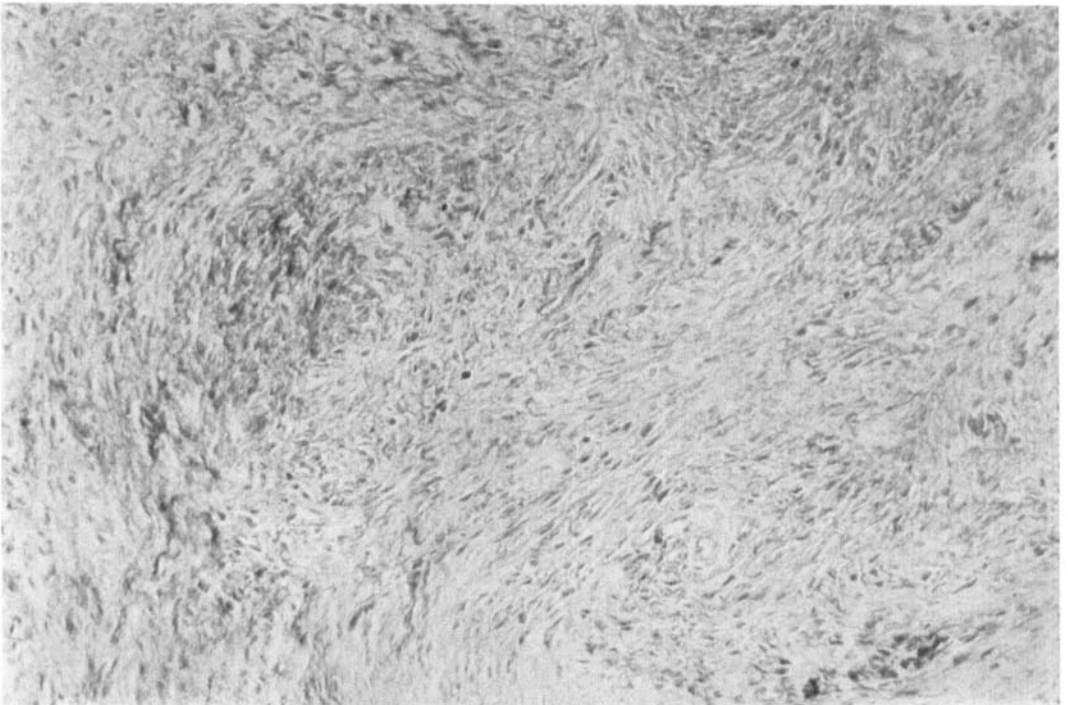


Figure 2b. Trichrome stain throws collagen fibrils into relief ( $\times 100$ ).

Table 1. Seventy-eight references dealing with 121 cases of desmoplastic fibroma of bone published in the period 1958–1982

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Table 2. Location of published cases of desmoplastic fibroma of bone

Mandibula	35
Femur	15
Humerus	12
Tibia	10
Radius	10
Ulna	1
Radius/ulna	2
Ilium	7
Os pubis	5
Scapula	4
Vertebrae	2
Fibula	2
Maxilla	5
Calcaneus	1
Clavicula	2
Ischium	1
Costae	2
Metacarpus	1
Metatarsus	1
Os sacrum	1
Os frontale	1
Unknown	1
Total	121

mittent pain was experienced in the following years.

In January 1980, the patient suffered a pathologic fracture of the distal part of the right humerus. A radiograph showed a fracture in the cystic area of the tumour, which in the meantime had recurred and progressed (Figure 3). In April, when the fracture had healed, the patient was operated on again. The compacta was found to be expanded and partly replaced by a membrane. The neoplastic tissue was rubbery and whitish gray and adhered to the endosteal surface. It was loosened by thorough curettage from the irregular cavity that extended distally to the subchondral bone. Microscopically, the tumour was on the whole unchanged since 1974, and it was classified as a desmoplastic fibroma. Radiographically, there was no sign of recurrence in February 1982 (Figure 4). Clinically, the patient was doing well in January 1983.

## Review of literature

Fifty-six of the cases in the literature were reported to have occurred in women and 56 in men. Seventy-five per cent of cases occurred in patients below 30 years of age. Symptoms such as pain and/or swelling (80 per cent) and patho-



Figure 3. Recurrence in 1980 before second operation.



Figure 4. Radiograph taken in 1982, 2 years after second operation.

logic fracture (13 per cent) dominated. Trismus because of tumour infiltration of the muscles of mastication occurred in four cases. In 40 per cent of cases, symptoms lasted for 1 year or more before diagnosis. The tumour is osteolytic and avascular. Radionuclide bone scans show increased uptake and CT-scans reveal soft tissue dissipation. Laboratory studies are normal. The most important differential diagnosis is the well-differentiated fibrosarcoma which mostly presents with a richer cellularity: larger irregular cells with plump and hyperchromatic nuclei and mitoses.

Treatment is reported in 101 cases, with 67 curettages and 29 resections. Forty per cent of these operations were supplemented with autologous grafting or an artificial prosthesis. In a few cases treatment consisted of irradiation, amputation, or chemotherapy. In 21 cases there is positive information about recurrence and in seven patients there were two or more recurrences. In 50 cases it was reported that there had been no recurrence. Recently Waddell & Gerner (1980) reported the effect of indomethacin and ascorbate on soft tissue desmoid in three cases is based on the assumption that the lesion is of an inflammatory rather than a neoplastic nature.

Because of the risk of recurrence, follow-up examinations should be frequent and be continued for several years.

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