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## DESMOPLASTIC FIBROMA OF BONE

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Cyst-like changes in roentgenograms of the skeleton are representative of numerous histo-pathological states. A probable diagnosis is all that can usually be made from x-rays. The last few years' research into bone tumours has permitted a greater differentiation in the assessment of the histo-morphological changes, with the recognition and clinical confirmation of new entities in the group known as cystic lesions of bone.

A *desmoplastic fibroma* was described by *Jaffe* (1958) as an unusual, benign tumour that appears on x-rays mostly as a cyst-like change in the skeleton. The diagnostic term is a reference to the great histological similarity of the tumour to the desmoid tumours in the abdominal muscles. Thus, the desmoplastic fibroma is greyish-white with a firm, fibrous consistency. Microscopically, it presents relatively few, small fibroblasts with abundant intercellular material rich in collagen fibres. There is no osteoid tissue. As a rule, x-rays show a central cyst-like trabeculated change in the metaphysis of a long bone. Occasionally, however, the lesion is more peripheral, in which case the cortex is thin and sometimes irregular.

The differential diagnosis may be difficult to make in respect of well-differentiated fibrosarcoma, chondromyxoid fibroma, non-ossifying fibroma and fibrous dysplasia. The last two states can generally be identified on the basis of the x-rays, but in the first two instances it may be difficult to differ the lesions from desmoplastic fibroma of bone. According to *Jaffe*, the histo-pathological differential diagnosis can be made on the following criteria: a well-differentiated fibrosarcoma is characterized by an increased richness of clearly polymorphic cells with large, plump nuclei. The chondroid and myxoid type of tissue in a chondromyxoid fibroma is easy to distinguish from the

fibrous tissue in the desmoplastic fibroma. Non-ossifying fibromas contain giant cells as well as foam cells, whereas desmoplastic fibromas do not. Finally, the tissue in fibrous dysplasia has regions with metaplastic ossification, whereas there is no osteoid tissue in a desmoplastic fibroma.

Jaffe based his account on five cases with an age range of 9–40 years. Three of the tumours were found in the tibia, the other two in the femur and scapula respectively. A few more cases of desmoplastic fibroma have been published since Jaffe's report, with the same age distribution but a variety of locations. The tumour appears to be unusual and we have only found 15 cases in the literature to date (cf. Table 1).

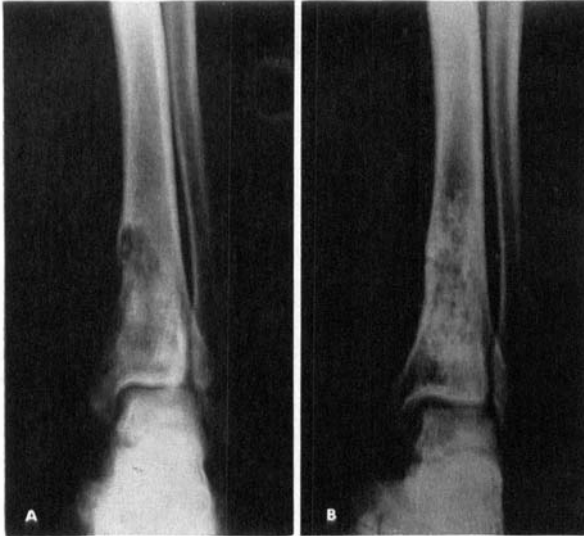
A study of our cases of fibrous lesions of bone disclosed 9 cases that represent the entity desmoplastic fibroma according to Jaffe's criteria. These cases are presented in some detail below.

*Table 1. References to 15 previously published cases of desmoplastic fibroma of bone.*

Author	Year	No. of cases
Jaffe	1958	5
Whitesides & Ackerman	1960	3
Scheer & Kuhlman	1963	1
Cohen & Goldenberg	1965	2
Godinho et al.	1967	1
Dahlin	1967	3
Total		15

#### OWN MATERIAL

*Csae 1* I.A., female, 18 years of age at first admission. For 6 months slowly increasing, ultimately severe pains above the right ankle. X-ray examination showed an osteolytic process situated somewhat eccentrically in the distal metaphysis and epiphysis of the right tibia. The cortex was irregular on the medial side (Figure 1A). Osteosarcoma was thought to be the probable diagnosis on this occasion. Amputation was suggested but the patient refused and was remitted to this clinic. At two operations, firm fibrous tissue was scraped out of the lesion. The first exploration showed that the process had a distinct but soft cortical wall and had not penetrated into the soft tissues. A histo-pathological examination showed a tissue poor in cells and with abundant intercellular material rich in collagen fibres; no signs of malignancy; diagnosis at present follow-up: desmoplastic fibroma. An X-ray check-up 4 years after the operations (Figure 1B) showed a somewhat irregular osseous structure with a well delimited outer cortical contour. Both then and subsequently,



*Figure 1. A. Desmoplastic fibroma in the distal tibia showing irregular osteolytic destructions. B. The same case 4 years after curettage now showing a clear cortical margin and a normalized though irregularly mineralized osseous structure.*

the patient was entirely free from discomfort, had no pains and normal mobility in the ankle joint. Observation time 16 years.

*Case 2.* U.R., female, 30 years of age at first admission. Treated with brachy-radium inserts for a cancer of colli uteri stage 1. Pains in the right half of the pelvis 8 months later led to an X-ray examination, which showed an osteolytic destruction close to the iliosacral joint in the right ilium. This was taken to be a metastasis from the uterine cancer and X-ray treatment was started. The diagnosis was, however, questioned and an operation was performed at which firm, rind-like fibrous tissue was scraped from the process in the right ilium. The tumour had not invaded the soft tissues. A histo-pathological examination showed hyalinised fibrous tissue poor in cells and containing fibroblasts with small, round nuclei. No signs of primary malignant or metastatic cancerous changes. Diagnosis at present follow-up: desmoplastic fibroma. The patient's symptoms disappeared after the operative treatment and she has subsequently remained entirely free from discomfort. Nor has any recurrence of the gynecological tumour been noted. Observation time 12 years.

*Case 3.* L.P., male, 15 years of age at first admission. For 6 months, pains in the left knee on movement. An X-ray examination showed a cystic change, almost the size of a walnut, located eccentrically in the distal femoral metaphysis, where it was expanding at the expense of the cortex. At operation, intraosseous firm fibrous tissue was removed. Microscopically, this comprised coarse bundles of hyaline connective tissue, poor in cells and without atypical cells. Diagnosis at present follow-up: desmoplastic fibroma. At a clinical examination in 1968, the patient was

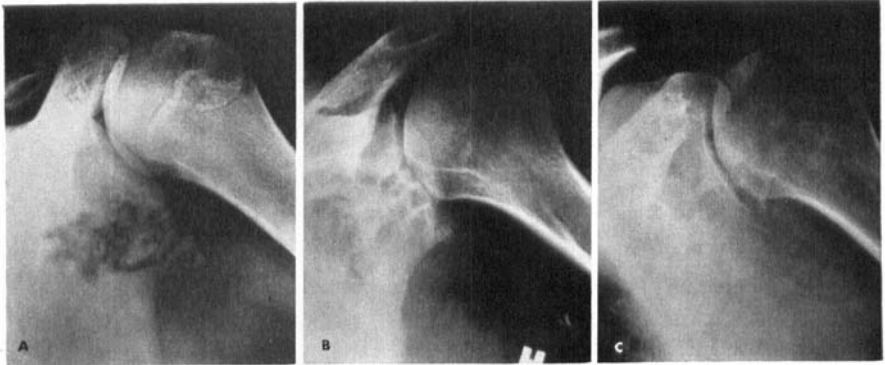


*Figure 2. Photomicrograph of desmoplastic fibroma. Bundles of dense fibrillary cells with little pleomorphism ( $\times 400$ ).*

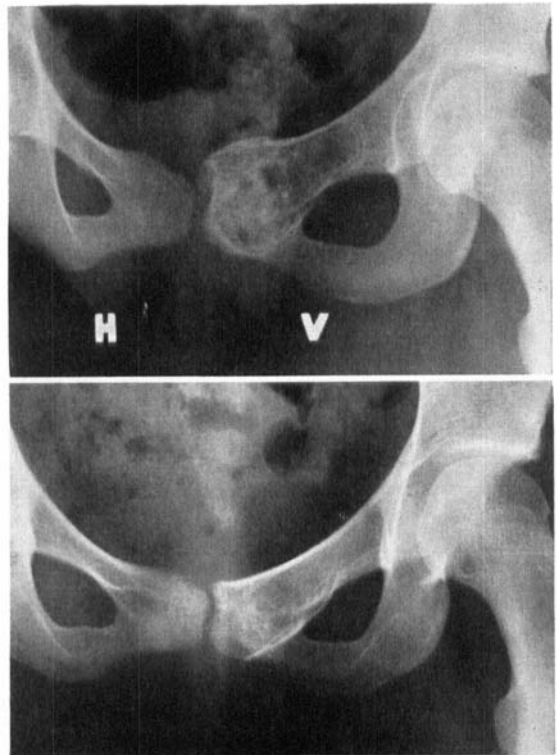
free from symptoms and roentgenograms showed that the process had healed practically without a trace. Observation time 12 years.

**Case 4.** R.A., male, 16 years of age at first admission. For 1 year, gradually increasing pain and loss of mobility in the right shoulder. An X-ray examination revealed osteolytic irregular destruction in the neck of the right scapula. At operation 1950, firm rind-like material was removed from intraosseous location in collum scapulae. A histological examination showed fibrous tissue poor in cells and rich in fibrils (Figure 2). No malignant structures were found. The cavities were filled without autologous bone chips (Figure 3 A). The symptoms recurred 10 years later and curettage was undertaken in 1960 and 1961 because X-rays suggested that the process might be progressive (Figure 3 B). The microscopic picture was unchanged. Diagnosis at present follow-up: desmoplastic fibroma. At clinical examination in 1968 the patient still had pains in the right shoulder and impaired mobility (abduction  $20^\circ$ , forward elevation  $30^\circ$ , fixed inward rotation  $45^\circ$ ). An X-ray examination showed small cysts in collum scapulae, otherwise normal osseous structure, and a moderate arthrosis in the humero-scapular joint (Figure 3 C). The patient's discomfort has obliged him to change to a lighter occupation. Observation time 18 years.

**Case 5.** B.A., male, 37 years of age at first admission. Pains in the right groin for 1 month led to an X-ray examination, which showed an irregular cystic invasion



*Figure 3. A. Curettage and bonegrafting of desmoplastic fibroma in collum scapulae (preoperative X-ray films not available). B. The same case 10 years later showing multiple cysts in the operated region and irregularities in part of the joint surface and the lateral margin of the scapula. C. The same case a further 8 years later. Only small remnants of the cysts and signs of slight osteoarthritis in the humero-scapular joint.*



*Figure 4. Desmoplastic fibroma in the superior pubic ramus (upper picture). The same case 5 years after curettage and bonegrafting showing an almost normal osseous structure (lower picture).*

of the inferior pubic ramus. A biopsy showed that the change was entirely intraosseous and contained firm, fibrous tissue. A histo-pathological examination showed a fibromatous tumour poor in cells, rich in collagen and without atypical cells. The diagnosis was desmoplastic fibroma. The tumour was treated with radical resection of the pubic bone from the symphysis to the ischial tuberosity and close to the acetabulum respectively. The defect in the floor of the pelvis was covered with plastic netting. Fistulas developed postoperatively from the region of the operation and persisted for 3 years. The patient was subsequently free from discomfort with no pains from the pelvic region or the leg. No signs of recurrence at X-ray check-up. Observation time 5 years.

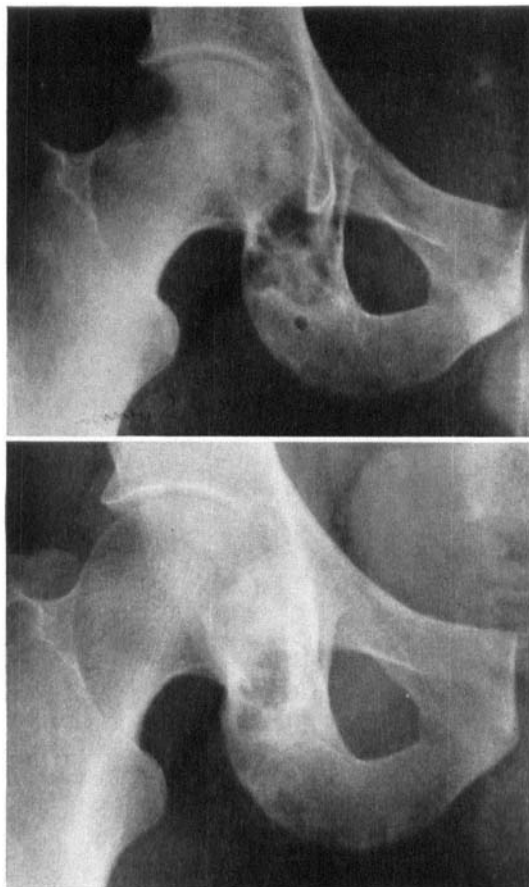
*Case 6.* E.K., female, 17 years of age at first admission. While skiing, the patient suddenly experienced a locking feeling and pains in the left groin without distinct trauma. An X-ray examination showed a fracture without dislocation through a well-defined cyst-like change in the superior pubic ramus (Figure 4). An operative exploration 1 month after the onset of symptoms showed that the lesion contained elastic, greyish-blue material that corresponded histologically to fibromatous tissue poor in cells and without atypical cells. In addition, however, there were traces of chondroid and osteoid tissue formation and occasional giant cells. Diagnosis: desmoplastic fibroma. After curettage, the cavity was filled with autologous bone chips. X-ray check-ups showed that these united well and in time the structure of the bone became practically normal (Figure 4). The patient's symptoms disappeared entirely. Observation time 5 years.

*Case 7.* J.H., female, 71 years of age at first admission. For 6 months, pains in right shoulder at rest and during movement. X-ray examination showed several, moderately trabeculated cysts in collum scapulae. At operation these were extirpated from a pale yellow, fibrous tissue. Microscopically, the lesion consisted of fibromatous tissue poor in cells. Diagnosis: desmoplastic fibroma. Postoperative X-ray check-ups showed that the structure of the bone gradually normalized. The patient became free from discomfort and practically normal mobility returned in the right shoulder joint. Observation time 4 years.

*Case 8.* S.E., female, 57 years of age at first admission. For 1 year, periodical pain in right groin at rest and on walking. X-ray examination disclosed a cyst-like change in the pubic bone just to the right of the symphysis. The cortex in the walls of the cavity had become thin and initial signs of calcification were observed in the surrounding soft tissues. A surgical exploration showed, however, that tumour was well delimited within the bone and had not invaded the soft tissues. The content of the lesion was extirpated and found to consist of firm, shiny fibromatous material. Histologically, this corresponded to coarse bundles of hyaline fibromatous tissue poor in cells. Diagnosis: desmoplastic fibroma. Postoperatively, the patient's symptoms disappeared. X-ray check-ups show that the bone structure is returning to normal, though there is sclerosis in the region of the operation. Observation time 3 years.

*Case 9.* E.S., male, 39 years of age at first admission. Pain at rest and on weight-bearing in the left groin for 6 months. An X-ray examination disclosed a somewhat

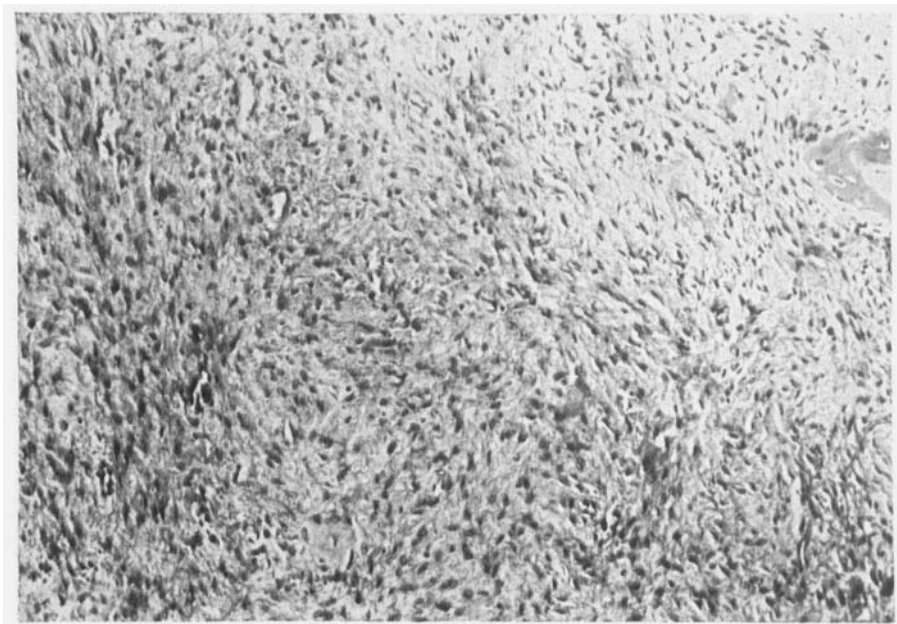
*Figure 5. Desmoplastic fibroma giving rise to an irregular osteolytic change in the superior ischial ramus (upper picture). The same case 2 years after curettage and bonegrafting showing sclerosis and also some thinner, trabeculated areas in the operated region (lower picture).*



irregular osteolytic change in the superior ischial ramus (Figure 5). At operation, a firm white fibrous mass was found within the bone and was extirpated, the resultant cavity being filled with heterologous bone. A histological examination showed well-differentiated fibrous tissue, moderately rich in cells and without cell polymorphism. Diagnosis: desmoplastic fibroma (Figure 6). X-ray check-ups showed normalization of the bone structure, though a thin trabeculated region remained (Figure 5). The patient is subjectively free from discomfort. Observation time 2 years.

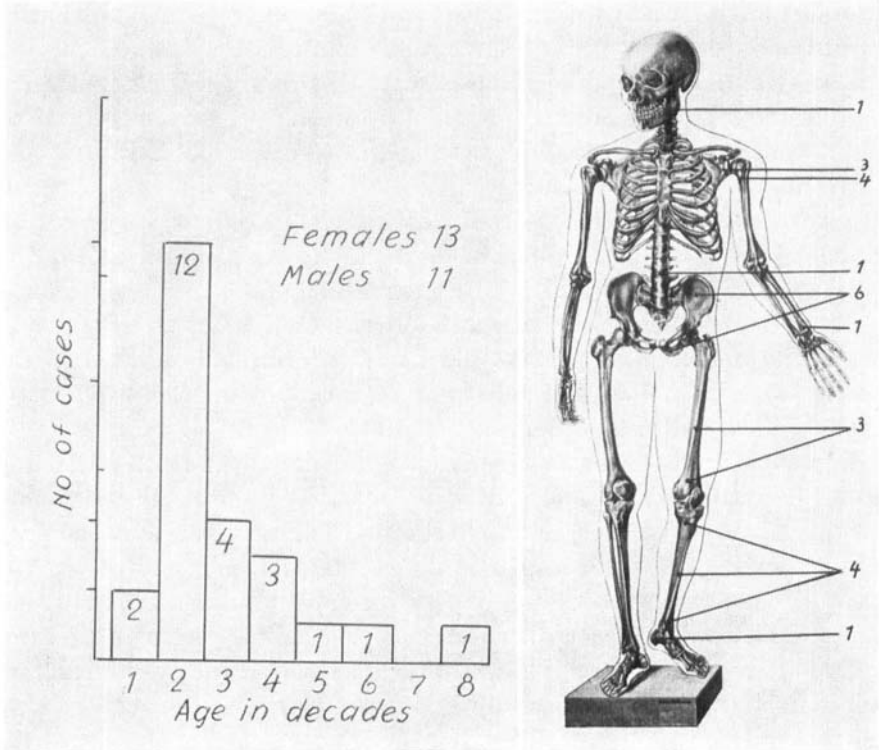
#### DISCUSSION

The diagnosis desmoplastic fibroma of bone in these nine cases appears to comply satisfactorily with Jaffe's criteria. In all cases, the lesion had an *intraosseous* location and the cortical surface was found to be intact at the first exploration. The content of the bone-lesion consisted of a



*Figure 6. Photomicrograph of desmoplastic fibroma. Homogeneous fibrous tissue with interlacing bundles of fibrillary cells ( $\times 150$ ).*

firm, fibrous tissue, generally greyish-white in appearance. The x-rays showed cystic or osteolytic changes, with multiple cavities and trabeculation in some cases. The extent of the destruction in a few cases suggested the possibility of malignancy. The histological examination consistently showed a fibre-rich, collagen tissue with relatively few cells. The fibroblasts were small and not atypical, so that well-differentiated fibrosarcoma could be ruled out in all cases. The long observation times—a range of 5–18 years for six of the cases—also suggest that the processes were not malignant. The absence of myxoid and chondroid tissue ruled out a diagnosis of chondromyxoid fibroma. In only one case (Case 3) was there reason to consider a diagnosis of non-ossifying fibroma. The x-rays showed an eccentric lesion in the femur, but the external wall was considerably thinner and bulged more than is usually the case in non-ossifying fibroma. The histological picture showed the coarse fibrillar bundles that are typical of desmoplastic fibroma in contrast to the looser connective tissue in non-ossifying fibroma. Osteoid tissue was found in Case 6, which suggested the possibility of a fibrous dysplasia. In this case, however, the biopsy was performed 1 month after a pathological fracture. The ossification



**Figure 7.** Distribution of 24 desmoplastic fibroma of bone (the 9 cases of the present series being added to the 15 previously published) by sex, age and localization.

was therefore taken to be a part of the current callus formation and the predominantly fibrous character of the lesion justified its classification as a desmoplastic fibroma.

Desmoplastic fibroma of bone appears to be a rare tumour. Dahlin, for instance, reported only 3 cases out of 3987 bone tumours. The 9 cases reported here and the 15 published previously add up to only 24 known cases of desmoplastic fibroma of bone. In view of the low incidence, it seems justifiable to draw certain general conclusions even though the number of cases is small.

As indicated by Figure 7, the tumour may occur at all ages, though the second decade predominates. More than half of all the cases were diagnosed in the period 10–30 years of age. There is no sex difference in the distribution of the tumours. The lesion may be located to a long bone as well as to the flat bones. It is worth noting that the tumour was in the pelvic bones or the scapula in 7 of the 9 cases reported here,

whereas only 2 cases with this location had been reported previously (Jaffe 1 case scapula, Whitesides & Ackerman 1 case ilium).

The symptoms are not characteristic, consisting of diffuse, moderate pain in the region of the tumour both at rest and on movement and/or weight-bearing. There may be local pains on palpation but as a rule there is no palpable resistance.

X-rays of desmoplastic fibroma in long bones show an osteolytic tumour with a central location in the metaphysis. The process may be somewhat transparent and is often trabeculated. The cortex becomes thinner as the tumour expands and sometimes appears so irregular as to suggest a malignant process. Desmoplastic fibromas located in the pelvic bones or the scapula are similar in principle but are often polycystic with a definitely sclerotic periphery.

Desmoplastic fibroma of bone is a benign tumour and can justifiably be treated with local surgery. Radical local resection of the lesion into healthy tissue is an acceptable alternative. This is often unnecessarily complicated, however, if the tumour is situated in *e.g.* the pelvis. Resection was employed in only one case in the present series. Five of the others were treated with curettage alone and the remaining three with curettage and bone-grafting. Both methods result in satisfactory bony union although the x-rays do not always show a completely normal osseous structure. This may indicate that regeneration of the bone occurs spontaneously once the fibrous tissue has been removed. We therefore recommend that the cavity resulting from the curettage be filled with bone-chips in order to accelerate union.

Having verified these nine cases as desmoplastic fibroma of bone, it seems likely that the tumour is somewhat more common than was previously supposed. After Jaffe's fundamental work it should be possible to be more certain about identifying a fibrous tumour in the skeleton as a desmoplastic fibroma. One should be particularly alert as regards cases that have previously been classified as well-differentiated fibrosarcoma, particularly if they are reported to have healed. The differential diagnosis between these two benign and malignant processes respectively is of the utmost importance for the choice of therapy.

#### SUMMARY

Nine cases of desmoplastic fibroma of bone are reported. This is the largest series presented to date and it brings the total number of registered cases to 24. The diagnoses were made according to the cri-

teria developed by Jaffe. The cases are presented with special reference to the roentgenologic and histological findings and to the principles of treatment.

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