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Desmoplastic Fibroma of Bone

—Report of a Case and Review of the Literature—

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Desmoplastic fibroma of bone is a very rare tumor. Jaffe(1958) described the desmoplastic fibroma of bone as "an uncommon benign tumor composed of small fibroblasts in a setting of abundant intercellular material which tends to be rich in collagen fibers". Radiologically the lesion tends to present as a centrally located radiolucent lesion in a long bone with a trabeculated or honeycombed appearance.

By 1976 fifty cases have been described(Sugiura, 1976). The rarity of this lesion and recently developed histogenesis of this lesion through electron microscopy prompted us to report the following case.

REPORT OF A CASE

This 13 year-old boy was admittted to the Seoul National University Hospital because of a mass in the 7th rib on June 2, 1981. The mass was incidentally discovered during the routine chest x-ray examination for tuberculosis screening.

There wasn't any complaint related to the rib lesion. Physical examination revealed neither palpable mass nor tenderness. Radiologically it was a radiolucent expanding lesion. The cortex was markedly thinned but intact. All the laboratory findings were noncontributory. Under the impression of benign bone tumor, segmental resection of the rib including intercostal muscle was done.

The specimen was a segmental portion of a rib, measuring 6 cm in length and 1 cm in diameter. The rib showed fusiform expansion in the central portion, measuring 2 cm in diameter. Cut surfaces showed whitish to grayish yellow hard well circumscribed mass with thin but intact cortical bone.

Microscopically the tumor consisted of irregularly arranged small spindle-shaped cells with fibroblastic appearance and intervening wavy collagen fibers. The tumor replaced the entire sponge bone and eroded the overlying compact bone. However, the tumor did not break through the cortical bone and the periosteum. The lesion was peculiar with relatively prominent large irregular blood vessels among the dense fibrous lesion. Focal hemorrhage with giant cell reaction was noted. Mitotic figure was not encountered.

DISCUSSION

A review of the world literature revealed 51 cases recorded(Jaffe, 1958; Whitesides and Ackerman, 1960; Scheer and Kuhlman, 1963; Dahlin and Hoover, 1964; Itami, 1964; Randelli, 1964; Cohen and Goldenberg, 1965; Griffith and Irby, 1965; Dahlin, 1967; Godinho et al., 1967; Hardy and Lehrer, 1967; Rabhan and Rosai, 1968; Hinds et al., 1969; Hovinga and Ingenhoes, 1969; Nissan et al., 1971; Scudese, 1971; Spjut et al., 1971; Cunningham et al., 1975; Nussbaum et al., 1976; Sugiura, 1976; Schenker and Kleinert, 1977; Dahlin, 1978; Huvos, 1979; Lagace et al., 1979; Mirra, 1980),

Sugiura (1976) collected 50 cases altogether in the world literature as of 1976. But most of Japanese articles cited by Sugiura (1976) were not available for review. The fifty-two patients including our case consisted of twenty-seven males, twenty-two females, and three patients whose sex was not recorded. Therefore, there is no significant sex difference.

The site distribution of fifty-two tumors are as follows(Table 1): the mandible (17.3%) is the most frequent site followed by femur (11.5%), humerus (11.5%), tibia (9.6%), and radius (9.6%) in decreasing order of frequency. The lesion tends to involve mandible, pelvic bone and long tubular bones.

The tumor occurs in patients with the age of 1 8/12~71 years, most frequently in the second decade(Table 2). Eighty-one per cent of the lesion occurs below 30 years of age.

Table 1. Site Distribution of 52 cases of Desmoplastic Fibroma

Site	No of Cases	Site	No of Cases
Femur	6(11.5%)	Clavicle	2(3.8%)
Humerus	6(11.5%)	Vertebra	3(5.8%)
Tibia	5(9.6%)	Calcaneus	1(1.9%)
Radius	5(9.6%)	Rib	1(1.9%)
Mandible	9(17.3%)	Metacarpal	1(1.9%)
Ilium	4(7.6%)	Scapula	3(5.8%)
Pubis	4(7.6%)	Skull	1(1.9%)
Ischium	1(1.9%)		

The roentgenograms of this neoplasm show cystic or osteolytic expanding lesion. When it occurs in long bone, the lesion affects the metaphysis or both ends of the shaft. Rarely the lesion extends to epiphysis as illustrated in two cases of Whitesides et al (1960), a case of Godinho's, a case of Nilsonne and Gothlin's (1969), and in a case of Lagace's (1969). Three cases involving midshaft of bone were reported (one case of Jaffe's cases, 1958; Cohen and Goldenberg, 1965; Scudese, 1971).

Table 2. Age Distristribution of Desmoplastic Fibroma

Age (yrs)	Male	Female	total	%
0~ 9	4	6	10	20.8
10~19	14	9	23	47.9
20~29	4	2	6	12.5
30~39	4	2	6	12.5
40~49		1	1	2.1
50~59		1	1	2.1
60~69				
70~79		1	1	2.1
Total	26	22	48	100

Usually the desmoplastic fibroma is centrally located but it may be eccentric as illustrated in one of five cases described by Jaffe(1958) and in case 3 by Nilsonne and Gothlin(1969). Both cases showed eccentric lesion in the femur, but the external wall was considerably thinner and bulged much more than is usually the case in nonossifying fibroma.

Radiographically, the expanding osteolytic lesion, with cystic cavities and trabeculation in some cases, may simulate aneurysmal bone cyst, chondromyxoid fibroma, fibrous dysplasia, solitary bone cyst, giant cell tumor, hemangioma, and eosinophilic granuloma. Extensive destruction with permeating border may suggest a metastatic carcinoma of thyroid or kidney (Whitesides, 1960) or osteosarcoma (Nilsonne and Gothlin, 1969).

Desmoplastic fibromas located in pelvic bones or the scapula are often polycystic with a definitely sclerotic border.

The clinical complaints of the patients are not specific as indicated in Table 3. The usual complaints are diffuse, moderate pain (64.9%) in the region affected. About one third of the patients complained of local swelling or palpable mass. Six out of 37 patients were admitted

Table 3. Complaints of 37 Cases of Desomoplastic Fibroma*

Complaints	No of Cases(%)
Pain	24 (64.9)
Limp	1 (2.7)
Swelling	11 (29.7)
Fracture	6 (16.2)
Disability	1 (2.7)
Discomfort	2 (5.4)
Inability to open mouth	1 (2.7)
Locking feeling	1 (2.7)
Loss of mobility	1 (2.7)

*Include two cases discovered incidentally.

because of fracture. The lesion presented here was discovered incidentally. There was another incidental lesion in the world literaturé (Jaffe, 1958).

The duration of the complaints before treatment was known for 37 of 52 patients (Table 4). The period ranged from one day (fracture) to 8 years. The complaints had been present for less than 6 months in 22(64.7%) of 34 patients, but in 12 patients (35.3%) the complaints had been present for more than 6 months..

The relatively high recurrence rate suggests that the tumor is biologically "borderline" as indicated in table 5.

There were 13 recurrences among 36 cases in which follow-up data were recorded. Healing occurred after amputation in 2 of 13 recurrences. There was high recurrence rate among those patients on whom curettage, curettage and bone graft, or simple excision was performed. But there was not any single recurrence among those groups in which wide local excision was done. It is becoming apparent that curettage or simple excision does not eradicate all the tumor tissue. Therefore, the lesions should be treated by wide local excision.

Grossly the lesion was usually whitish or grayish or grayish white, of firm or rubbery consistency. It looked just like a dense fibrous tissue. Sometimes occasional small cavities were found on cut surfaces (Rabhan and Rosai, 1968) Microscopically the lesion was similar to desmoid tumor of soft tissue. The tumor consisted of spindle cells of fibroblastic appearance surrounded by abundant wavy collagen fibers.

Table 4. Duration of Complaints of Desmoplastic Fibroma

Duration	No of Case(%)
Less than 1 month	10(29.4)
1 month to 6 months	12(35.3)
6 months to 1 year	8(23.5)
1 year to 5 years	3(8.8)
More than 5 years	1(2.9)

The cellularity varied from case to case. According to Rabhan et al. (1968) there appeared to be a correlation between the cellularity of the lesion and the incidence of recurrence.

From the morphological point of view, there are five lesions which occasionally could pose problem in the differential diagnosis. They are well differentiated fibrosarcoma, non-ossifying fibroma, chondromyxoid fibroma, fibrous dysplasia, and soft-tissue desmoid. In fibrosarcoma, the tumor cells are relatively large and plump with mitosis in areas, while small fibroblasts with rather inconspicuous nuclei and abundant collagen formation characterize desmoplastic fibroma.

Table 5. Recurrence Rate according to Treatment Modality

Treatment	No of Cases	Recurrence(%)
Curettage	7	2(28.6)
Curettage and bone graft	10	5(50)
Simple excision	8	6(75)
Wide local excision	11	0(0)
Total	36	13(36.1)

Many giant cells in the background of whorled bundles of small spindle-shaped cells and occasional xanthomatous cells are hallmark of non-ossifying fibroma, posing little problem, if any, in delineating from desmoplastic fibroma.

Pseudolobulation with myxoid and fibrous component in chondromyxoid fibroma contrasts with the findings in desmoplastic fibroma. The lack of osseous metaplasia serves to aid in differentation from fibrous dysplasia. The differentiation between desmoplastic fibroma and soft-tissue desmoid depends solely on clinical and x-ray findings, because the histopathologic findings are exactly the same.

Usually desomplastic fibroma of bone is well defined by expanding cortical shell, but sometimes extend to surrounding soft tissues. The soft-tissue desmoid manifests as a soft tissue mass either intramuscular or periosteal. Occasionally soft-tissue desmoid may be attached to underlying bone and erode the cortical bone.

Desmoplastic fibroma of bone is rare. Only four cases were found among 6,221 pathologically verified bone tumors in the files of the Mayo clinic(Dahlin, 1978).

Usually the lesion presented as a problem in orthopedic and dental clinic. But present case is peculiar in that the patient was presented to chest surgeon because of mass in the rib. The lesion was discovered incidentally through routine chest x-ray.

Concerning the histogenesis, Jaffe (1958) suggested fibroblastic origin. In his original description, Jaffe (1958) states "—the name desmoplastic fibroma" for the lesion, —primarily to indicate the densely fibrous character of the lesional tissue, which makes it resemble the familiar desmoid tumor of the abdominal wall". According to Sugiura (1976) the tumor was composed solely of fibroblasts. But Lagace et al (1979). suggested myofibroblastic origin. They found that the principal cellular component was myofibroblast, while a limited number of fibroblasts and primitive mesenchymal cells were also noted. Through electron microscopy, they demonstrated many cells which have numerous bundles of cytoplasmic filaments with zone of osmiophilic condensations, nuclear deformation, pinocytic vesicles, intercellular connections and basement membrane material. These findings are distinct from conventional fibroblasts. The presence of many myofibroblasts is not surprising for they are a common feature of fibromatoses.

In short, desmoplastic fibroma of bone is a rare benign fibrous connective tissue tumor of probable myofibroblastic origin. The lesion is usually sparsely cellular with spindle-shaped nuclei and characterized by abundant collagen formation. Although the lesion is histologically benign, it's biologically "borderline" with frequent recurrences, indicating desirability of wide local excision as a treatment of choice.

SUMMARY

A case of desmoplastic fibroma of the 7th rib in a 13 year-old boy was presented. The lesion presented incidentally on the routine chest x-ray as a expanding osteolytic lesion.

Critical review of the world literature indicates that desmoplastic fibroma of bone is a distinct benign fibrous connective tissue tumor of probable myofibroblastic origin with biologically "borderline" course, necessitating a wide local excision as treatment of choice.

=국문초록=

골의 건종양 섬유종

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13세된 남아의 좌측 제 7 늑골에 발생한 건종양 섬유

종을 관찰하고 이를 기술하였다. 결핵검진시 흉부 X-선상 우연히 발견되었으며 제 7 늑골의 팽대된 골조직 융해성 병변으로 팽대된 골 변연부는 잘 유지되어 있었다. 세계문헌상 최초의 늑골병변으로 흉부외과에서 수술을 받았다. 본례는 한국문헌상 최초의 골의 건종양 섬유종으로 사료되었다.

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LEGENDS FOR FIGURES

Fig. 1. Roentgenogram of the chest showing expanding osteolytic lesion of 7th rib.

Fig. 2. Roentgenogram of the expanding osteolytic lesion of 7th rib showing eccentric position, its uniformly rarefied appearance, as well as thinning and expansion of the overlying cortex, which is still intact.

Fig. 3. Cut surface of the desmoplastic fibroma of bone showing gray white hard tumor with intact thin cortex.

Fig. 4. Photomicrograph of the desmoplastic fibroma of bone, showing sparsely cellular tumor. H. & E. ×200.

Fig. 5. Low power view of desmoplastic fibroma of bone, showing small spindle-shaped cells with abundant deposition of collagen fibers. H. & E. ×200.

Fig. 6. High power view of cellular area of desmoplastic fibroma of bone, showing spindle cell of fibroblastic appearance with apparent wavy collagen formation. H. & E. ×430.


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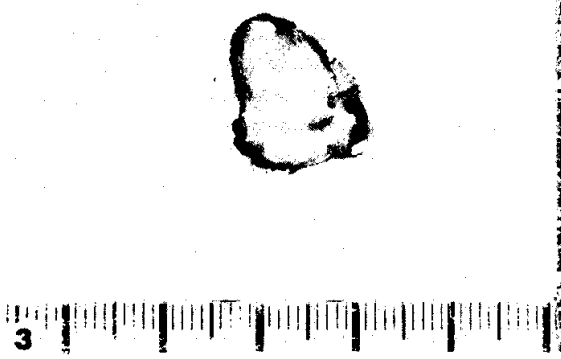
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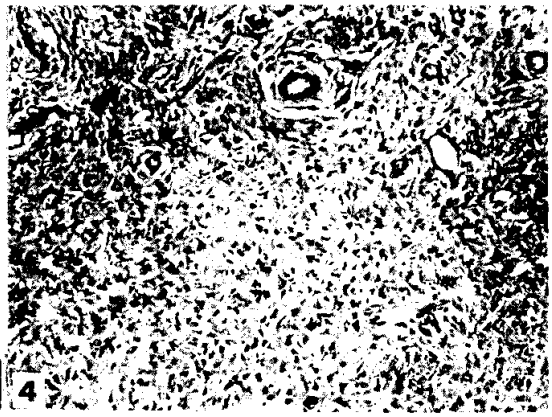
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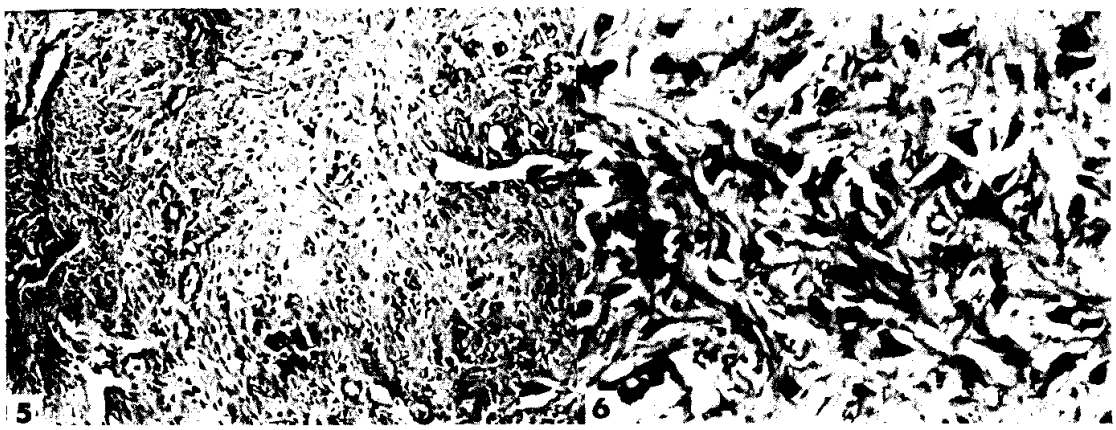
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