

Ossifying Fibroma

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Our experience includes seven cases of ossifying fibroma. The condition also appears in the literature under diagnostic names such as congenital fibrous dysplasia, congenital osteitis fibrosa, congenital fibrous defect of the tibia, and osteofibrous dysplasia of the tibia and fibula. The lesions develop in childhood and are located in the diaphysis of the tibia, or fibula.

Of seven patients, we performed wide excision with freevascularized fibular graft in five cases, wide resection of the distal one-third of the fibula in one case, and curettage and bone graft in one case. Two of the patients who had wide excision with free vascularized fibular graft had recurrence. One case of recurrence occurred where incomplete wide excision with free-vascularized fibular graft was performed because the lesion was too close to the distal epiphysis of the tibia. One of the patients who had curettage and bone graft also had recurrence. It was concluded that children who have an ossifying fibroma requiring surgery can safely be treated with wide excision with or without free-vascularized fibular graft.

Key Word: Ossifying fibroma

Ossifying fibroma is not a well-recognized entity. It usually develops in the maxilla and mandible, and only rarely affects long bones. If the lesion is located in the long bones, it usually affects the tibia or fibula (Schonecker et al. 1982).

Ossifying fibroma seldom has even a moderate tendency to progress during childhood, but it recurs frequently after curettage or subperiosteal resection. Any progression of the lesion comes to an end after puberty. Thus, surgery should be delayed as long as possible. However, if the lesion is rapidly progressive, or if a patient has a repeated fracture, it would be necessary to resort to wide extraperiosteal resection (Campbell and Thomas 1982).

Our report includes a review of 7 cases. Twenty eight months was the average long-term followup in these cases.

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MATERIALS AND METHODS

Seven patients were treated between October 1979 and November 1988 at the Yonsei University College of Medicine. Six of the patients were less than ten years old and one patient was thirteen years old. There were five male and two female patients. Six of the lesions occurred in the tibia and one was in the fibula. Mild pain, limping gait, bony enlargement and bowing deformity were the presenting symptoms (Table 1). All patients had extensive lesion.

Subperiosteal curettage and autogenous bone graft was the initial treatment in one of the patients. Of the other six patients, wide excision with free vascularized fibular graft was done in five patients, and wide resection of the distal one-third of the fibula was done in one patient (Table 1).

The patients who had excision with free vascularized fibular graft were immobilized with a long-leg cast. Full-weight bearing was permitted 15 months after operation.

Table 1. Clinical data on seven patients with long term follow-up

Case	Initial presentation			Bone involved	Initial operation	Recurrence	Subsequent operation	Age (Yrs)	Last follow-up	
	Age (Yrs)	Sex	Date	Complaint					Date	Remark
1	5	M	1978	Pathologic Fracture	Tibia	Curettage & Bone graft	2 Times	Curettage & Bone graft	7, 8	Lost
2	6	M	1985	Painful Swelling	Tibia	Wide excision & F.V.F.G.*	NO	-	-	1987 Good Healing
3	9	M	1986	Palpable Mass	Fibula	Wide resection	NO	-	-	1988 Good Recurrence
4	13	M	1986	Anterior Bowing	Tibia	Wide excision & F.V.F.G.*	NO	-	-	1989 Good Healing
5	6	M	1986	Anterolateral Bowing	Tibia	Wide excision & F.V.F.G.*	3 times	Curettage & Bone graft	7, 8, 9	1989 Good Healing
6	4	M	1988	Palpable Mass	Tibia	Wide excision & F.V.F.G.*	Once	Curettage & Bone graft	5	1989 Good Healing
7	8	F	1988	Limping Gait	Tibia	Wide excision	NO	-	-	1988 Good Healing

* Free Vascularized Fibular Graft

RESULTS

The patient who had curettage and autogenous bone graft had two recurrences. Subsequent curettage and allogenic bone grafts were performed. Follow-up was not possible three years after the initial operation.

The patient with a fibular lesion had wide resection of the distal one third of the fibula. There was no recurrence in this case.

Of the five patients who had wide excision with free-vascularized fibular graft, two patients had recurrence. One case of recurrence occurred where incomplete wide excision with free-vascularized fibular graft was performed because the lesion was too close to the distal epiphysis of the tibia. In this case recurrence occurred three times. Another case of recurrence occurred where complete wide excision with free-vascularized fibular graft was performed, but there was only one recurrence. Two other patients with recurrent tumor were treated by subsequent curettage and bone graft.

To illustrate the wide excision with free-vascularized fibular graft, three of these patients will be discussed in further detail.

Remarkably, all patients in this study had full range of motion in the knee and ankle joints after surgery.

Case 1.

A thirteen-year-old girl was seen on August 20, 1986 with an eight year history of an anterior bowing deformity of the left tibia. Radiographs revealed an extensive osteolytic lesion and anterior bowing of the left tibia (Fig. 1-A). On August 27, 1986, the patient underwent a wide excision of the left tibia (18 cm) with free-vascularized fibular graft (Fig. 1-B). A gross specimen showed that the external surface was bluish gray and mass which contained numerous minute calcifications. Histologically, the bony spicules which contain well developed osteocytes were rimmed by osteoblasts (Fig. 1-C). In July 1987, the patient had a slip-down injury, and a fracture occurred in the grafted fibula (Fig. 1-D).

She was treated with a long leg cast, and the fracture completely healed after 5 months (Fig. 1-E). She remained free of disease for thirty-three months after operation, and radiographs showed the complete union of the grafted bone and hypertrophy of the grafted fibula (Fig. 1-F).

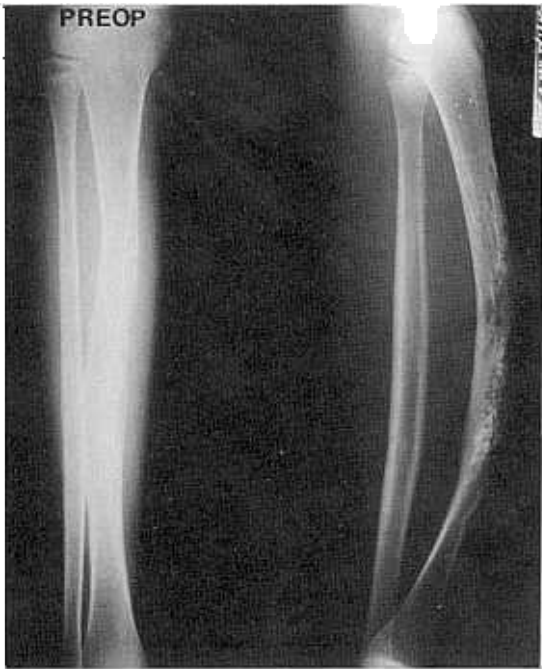


Fig. 1-A. Initial radiograph shows the extensive osteolytic lesion and anterior bowing on the left tibia.



Fig. 1-B. Radiographs after wide excision and free-vascularized fibular graft.

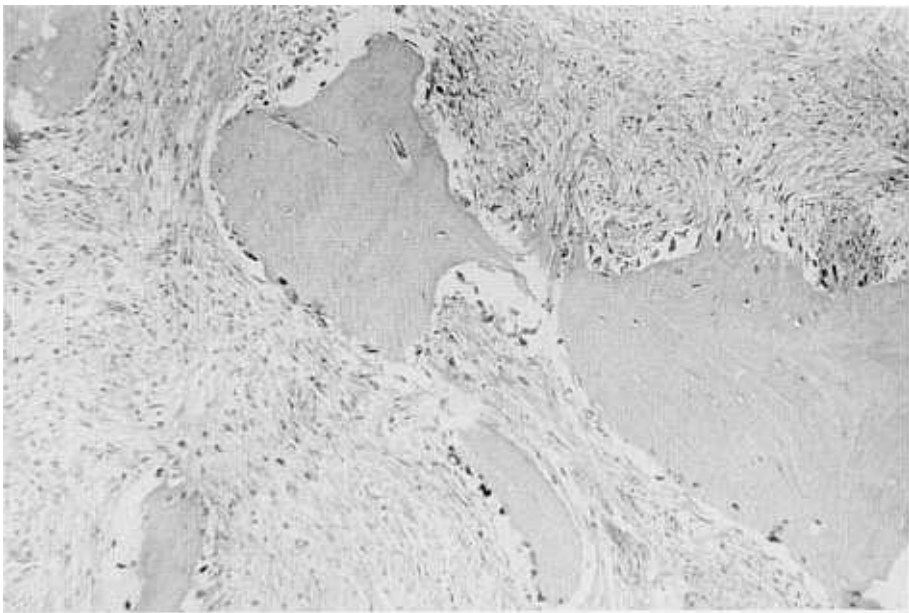


Fig. 1-C. Photomicrograph of histologic section of case 1 shows the bony spicules which contain well-developed osteocytes and are rimmed by osteoblasts (H-E, $\times 200$).



Fig. 1-D. Eleven months after operation, radiographs show a fracture line in the grafted fibula after slip-down injury.

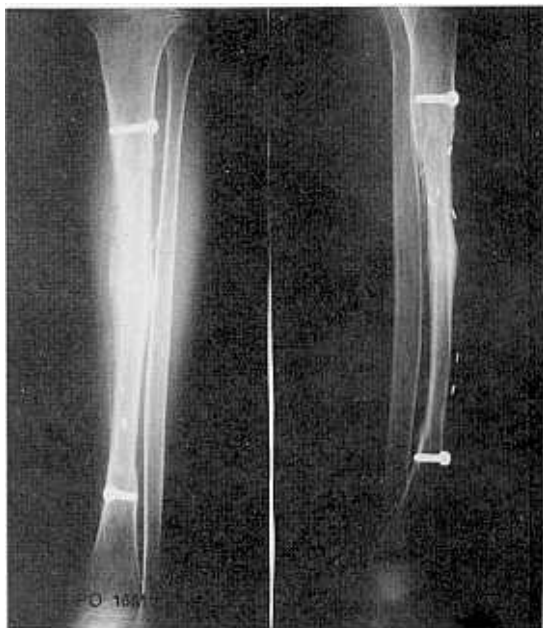


Fig. 1-E. Five months after fracture, radiographs show the complete union of the fracture.



Fig. 1-F. Thirty-three months after operation, radiographs show the complete union of the grafted bone and hypertrophy of the grafted fibula. There was no evidence of recurrence.

Case 2.

This six-year-old boy was seen on December 20, 1986 with an anterolateral bowing deformity of the left tibia since birth. The patient had been treated with curettage and allogenic bone graft twice at another hospital. Radiographs revealed an extensive osteolytic lesion on the proximal and distal portion of the left tibia. Anterolateral bowing is also present. There was also a pathologic fracture on the distal portion of the tibia (Fig. 2-A). On December 31, 1986 the patient underwent an incomplete wide excision with free-vascularized fibular graft because the lesion was too close to the distal epiphysis of the tibia (Fig. 2-B). A gross specimen showed that the outer surface was smooth and grayish brown, and an ill-defined, grayish white firm mass involved the intramedullary space and cortex. The histological appearance was similar to that of the previously described lesion (Fig. 2-C). In December 1987, the patient showed a cystic recurrence in the distal portion of the tibia and curettage and an autologous bone graft was performed. In September 1988, again cystic changes occurred in the proximal and distal portion of the left tibia. The patient un-



Fig. 2-A. Initial radiographs show the extensive osteolytic lesion on the proximal and distal portion and anterolateral bowing of the left tibia.



Fig. 2-B. Radiographs after wide excision and free-vascularized fibular graft.

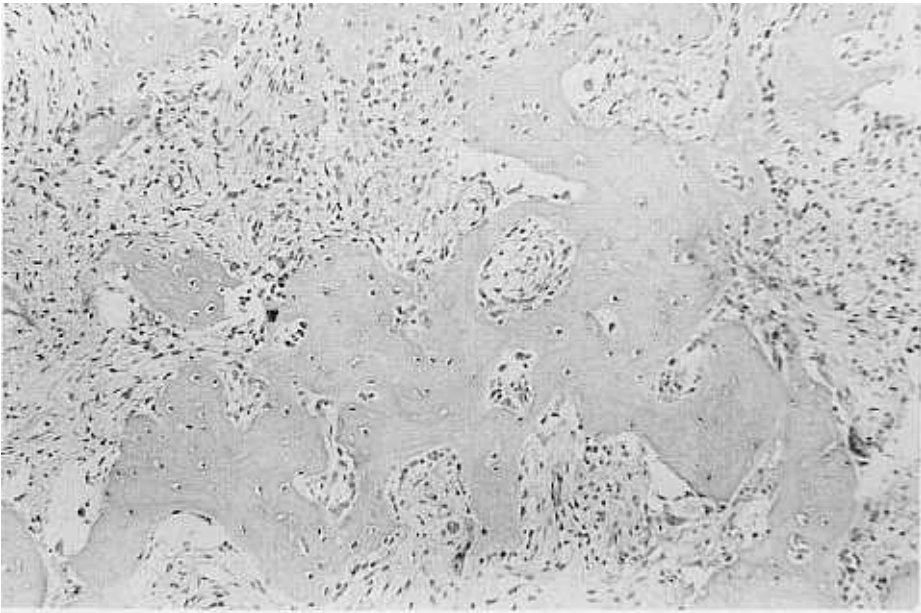


Fig. 2-C. Photomicrograph of histologic section of case 2 shows the randomly dispersed mature bone trabeculae which are present within dense cellular fibrous tissue (H-E, $\times 200$).



Fig. 2-D. Thirty nine months after operation, the recur-red osteolytic lesion was noticed on the proximal and distal portion of the left tibia.



Fig. 3-A. Intial radiographs show the extensive osteolytic lesion and cortical thinning and bulging on the left tibia.



Fig. 2-E. Radiographs after curettage and allogenic bone graft.



Fig. 3-B. Radiographs after wide curettage and free-vascularized fibular graft.

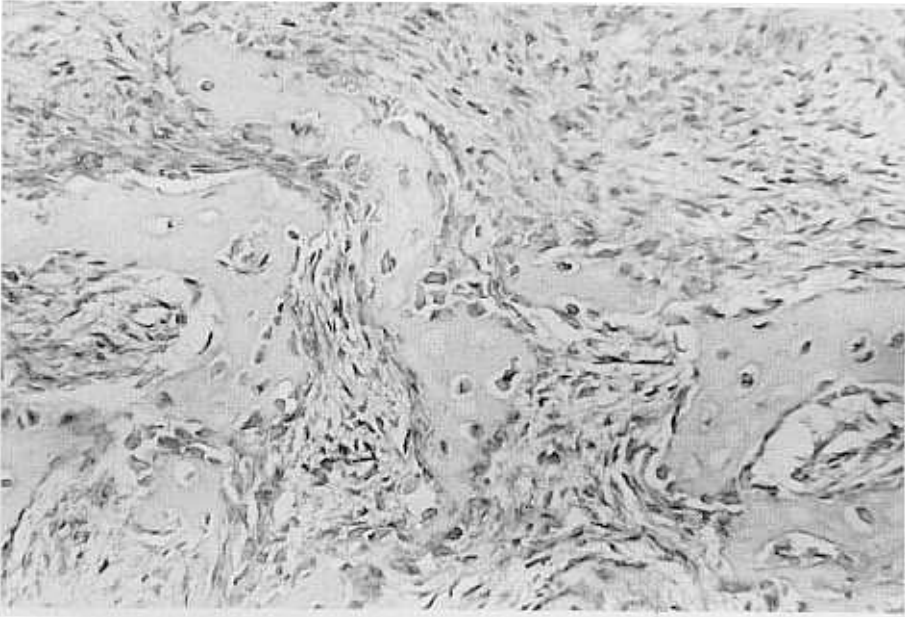


Fig. 3-C. Photomicrograph of histologic section of case 3 shows the occasional woven bone and osteoblasts rimming (H-E, $\times 200$).

derwent curettage and autoiliac bone graft. In May 1989, the lesion recurred once again on the proximal and distal portion of the left tibia. Curettage and allogenic bone graft were once again performed (Fig. 2-D, Fig. 2-E). In this patient, we could not perform complete wide excision because the lesion was too close to the distal epiphysis of the tibia.

Case 3.

A six-year-old boy was seen on March 8, 1985 with a one year history of painful swelling in the distal part of the left leg. Roentgenograms of the left tibia demonstrated an extensive osteolytic lesion and cortical thinning and bulging (Fig. 3-A). On March 20, 1985, a wide excision (14.5 cm) with free-vascularized fibular graft was performed (Fig. 3-B). Histological examination of the lesion revealed spicules of bone with a fibrous stroma. The bony spicules were rimmed by osteoblasts (Fig. 3-C). Seventeen months after this operation, there was no roentgenographic evidence of tumor recurrence (Fig. 3-D).



Fig. 3-D. Seventeen months after operation, radiographs show the complete union of the grafted bone and hypertrophy of the grafted fibula. There was no evidence of recurrence.

DISCUSSION

Ossifying fibroma was reported in 1921 by Franghenheim, who named the entity "congenital osteitis fibrosa". Aside from congenital fibrous dysplasia, the terms congenital fibrous dysplasia and congenital fibrous defect of the tibia have been used. In 1981, Campanacci and Laus reported thirty-five patients with as osteofibrous dysplasia of the tibia and fibula.

In 1966, Kempson reported two patients with ossifying fibroma of the long bones who had distinct clinical, histological and ultrastructural differences from monostotic fibrous dysplasia. Campanacci and Laus proposed the term osteofibrous dysplasia of the tibia and fibula for the entity because it was present exclusively in those bones. In our study, the lesion occurred in the tibia and fibula.

The pathogenesis of ossifying fibroma is not known. Kempson reported that the electron micrographic finding indicated both bone deposition and resorption occurring on the same spicules of bone. It was postulated that ossifying fibroma resulted from excessive resorption of bone with fibrous repair of the defect. The initial resorption might be secondary to defective bone and probably represent attempts at healing. Whatever the basic defect, the role of the periosteum seemed significant. The periosteum could be stimulation production of abnormal osteoblasts, or an excessive numbers of osteoclasts. Therefore, if surgery is indicated, wide excision involving the normal periosteum is mandatory (Campanacci and Laus 1981).

The symptoms almost always appear in the first decade of life, always before puberty. In our study, six patients were in the first decade and one patient was a thirteen-year-old. The lesion almost always occurs in the tibia or fibula, but occasionally in the ipsilateral tibia and fibula or bilaterally. Also it has been reported in the humerus of one patient (Schitter 1958). In this study, the lesion occurred in the tibia of six patients and in the fibula of one patient.

The most frequent presenting symptom is enlargement of the tibia, usually associated with slight or moderate anterior or anterolateral bowing. Pathologic fracture or pseudoarthrosis of the tibia or fibula rarely occurs (Campanacci and Laus 1981). In this study, enlargement or mass of the tibia was observed in three patients, anterior or anterolateral

bowing in two patients, limping gait in one patient, and pathologic fracture in two patients.

Radiographically, eccentric intracortical osteolysis is seen, and the external surface of the cortex may be moderately or severely expanded. The osteolysis is clearly margined by a band of sclerosis, and frequently there is narrowing of the medullary canal. Anterior or anterolateral bowing of the tibia is regularly associated with more extensive lesions. These radiographic features are so characteristic that diagnosis can be made, with confidence, from the radiographs alone, before or without histological confirmation. However, there is no radiographic criterion to predict the course of the lesion (Campanacci and Laus 1981).

Histological examination shows two fundamental characteristics. First, benign fibrous tissue with numerous irregularly shaped trabeculae of woven bone which are often rimmed by osteoblasts is seen. Second, a so-called zonal architecture has been described with a transition from sparse, immature trabeculae of woven bone in the center of the lesion to more mature trabeculae of lamellar bone near the cortex.

Adamantinoma, monostotic fibrous dysplasia and non-ossifying fibroma can be differentiated from ossifying fibroma clinically, radiographically and by the histological pattern. Adamantinoma seldom occurs in the first decade and a typical epithelioid component can be found in histological examination. Monostotic fibrous dysplasia usually involves the femur and the rib. It is usually discovered after the patient has reached the age of ten years. Its radiographic feature is usually intramedullary and the bowing of the tibia is not seen. Histologically, fibrous dysplasia has more cellular and less mature fibrous tissue, and usually shows only in woven bone, not a so-called zonal architecture. Non-ossifying fibroma usually involves the metaphysis, and histologically, there is no new bone formation except in its reparative phase (Marbel 1978; Park et al. 1978).

Ossifying fibroma usually progresses until ten years of age and recurs frequently after curettage or subperiosteal resection, but occasionally regresses spontaneously after puberty. Therefore, surgery should be delayed as long as possible. But if the lesion is rapidly progressive, if a patient has a repeated fracture, or when surgery is absolutely necessary to maintain the integrity of the involved extremity, it would be necessary to eradicate the lesion completely (Georgen et al. 1977). In this study, we performed wide excision with free-vascularized fibular graft in five patients and wide resection of the distal

one-third of the fibula in one patient. Two patients had recurrence. One case of recurrence occurred where incomplete wide excision with free vascularized fibular graft was performed because the lesion was too close to the distal epiphysis of the tibia.

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