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NON-OSTEOGENIC FIBROMA OF BONE

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Localized fibrous defects in the metaphysis of the long bones have attracted considerable interest through the past thirty years. The radiographic and histological appearances of these defects are known, but there is not agreement concerning their true nature, classification, or the terminology. On the basis of our present knowledge it is difficult to decide whether these fibrous defects make up a pathogenic entity or whether they are to be classified into two or more types. Certain criteria indicate that the latter would be more correct, and in accordance with this view non-osteogenic fibroma of bone may be defined as a special type of the localized fibrous defects which occur in the metaphysis of long bones in children and adolescents.

Little mention seems to have been made of non-osteogenic fibroma of bone in the Scandinavian literature. The object of this paper is to review the main features of the disease and to present a Norwegian material.

The names given to the disease and its assumed aetiology have varied. Phemister, who was the first to describe the fibrous defects (1929), believed that the lesions were caused by chronic, non-specific infection and named the disease chronic fibrous osteomyelitis. Phélip (1935) and Burma & Sinberg (1938) interpreted the condition as lipoid granulomatosis and introduced the terms juvenile cystic xanthomatous osteitis and solitary xanthomas of bone. Jaffe & Lichtenstein (1942) concluded that it was a benign tumour arising in the fibrous tissue of the bone marrow, that these particular fibromas were not associated with bony metaplasia of the proliferating fibrous tissue, and suggested that the disease be called non-osteogenic fibroma of bone. Later, it has been demonstrated (Sontag & Pyle 1941, Caffey 1955) that fibrous defects of the same histological appearance as Jaffe & Lichtenstein's non-osteogenic fibroma of bone and with a marked

tendency for spontaneous regression very often occur in the metaphysis in children. On this basis, Hatcher (1945), Maudsley (1956), and Compere & Coleman (1957) concluded that non-osteogenic fibroma of bone could not be separated from other fibrous metaphyseal defects, that the lesion could not be a neoplasm, and that very probably it was a developmental anomaly, possibly an intermittent disturbance of the process of ossification. This relationship between the common cortical metaphyseal defects and non-osteogenic fibroma of bone does not appear to be generally accepted. On the other hand, it seems to be agreed today that non-osteogenic fibroma of bone is not a neoplasm and that presumably the lesion represents fibrous changes following necrosis left by infarction (Robbins 1962).

The frequency of localized fibrous defects in the metaphysis of long bones was studied by Sontag & Pyle (1941). In periodical serial investigations of 200 otherwise healthy children they found that 54 per cent of the boys and 22 per cent of the girls had defects in the cortical layer of the juxta-genual metaphysis at some time or another between 2 and 18 years of age. A histological examination of these defects was not performed. All the defects subsided spontaneously within 29 months. Caffey (1955) demonstrated that X-ray examination of the knee in children aged 2–12 years disclosed cortical metaphyseal defects in 7–20 per cent. The last-mentioned author also found the defects to subside spontaneously within an average period of 4.4 years in the boys and 2.1 years in the girls. This author described the histological picture of the defects but did not mention how often a histological study was done. In his opinion, the cortical metaphyseal defects of his series are not identical with Jaffe & Lichtenstein's non-osteogenic fibroma of bone, but he admits that the histological appearances are similar. As the terminology and classification are not agreed upon, and as the localized fibrous defects are often silent, it is not possible to state the exact incidence of non-osteogenic fibroma of bone. According to clinical experience, however, it is much lower than that found for cortical metaphyseal defects, and this marked difference in the incidence is a strong argument in favour of separating the two conditions.

As a rule, subjective symptoms are not outstanding, and in half the cases entirely absent. In such cases the diagnosis is incidental, made after X-ray examination for traumas. In the other half, the symptoms consist of mild pain and discomfort in the metaphyseal region or adjacent joint. There may also be swelling of the soft tissues overlying

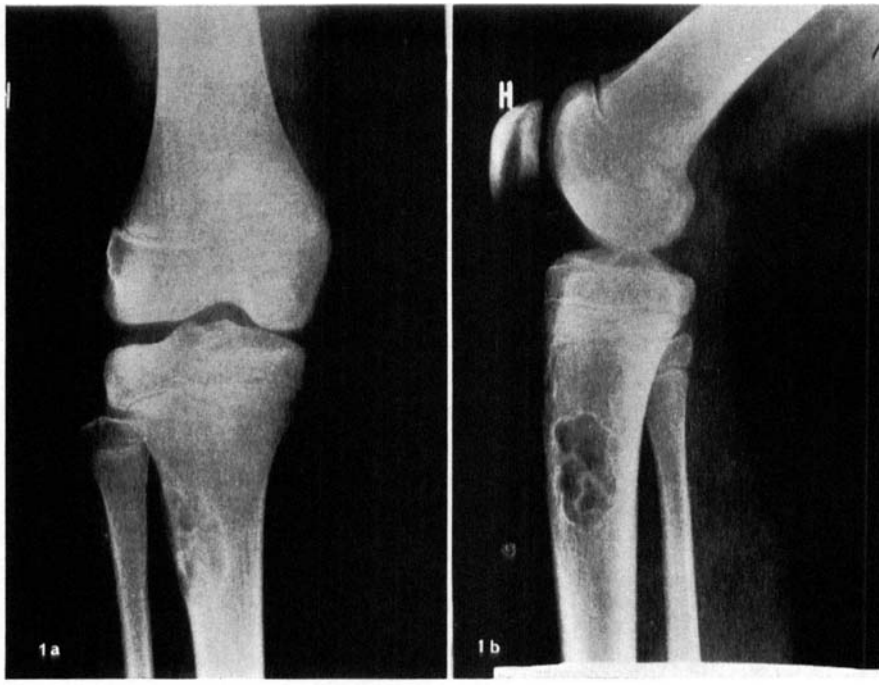


Figure 1 a and b. 14–year–old girl. No symptoms prior to trauma. Histological examination revealed non-osteogenic fibroma of bone.

the affected bony area. Not infrequently, a pathological fracture is what leads the patient to a doctor.

The X-ray appearances of non-osteogenic fibroma of bone are characteristic, but may vary a bit from patient to patient, presumably according to the age of the lesion. It presents a well-defined area of cyst-like translucency in the metaphysis of the long bones. Fresh lesions are localized in the vicinity of the epiphyseal line, older lesions at a somewhat greater distance. In the large bones (femur, tibia, humerus) (Figure 1), the lesions are of an eccentric site, having a long axis parallel with that of the bone. In the smaller bones (fibula, radius, ulna), on the other hand, the defect is central, and the bones show a fusiform bulging at the site of the defect. As a rule, the defects are multilocular, but they may be oval or almond-shaped. Their maximum diameter seldom exceeds 5 cm. A periosteal reaction is never present unless there is a fracture through the defect. Against the normal cancellous bone the defects are delimited by a distinct

Table 1.

Case	Age sex	Site	Symptoms	Treatment	Histology
1	17 ♂	Distally in R. tibia	None	Curettage	Non-osteogenic fibroma of bone
2	14 ♂	Distally in R. femur	None	Curettage + bone grafting	Fibrous defect (non-osteogenic fibroma of bone)
3	9 ♀	Distally in R. tibia	Patho-logical fracture	Curettage + bone grafting	Probably non-osteogenic fibroma of bone
4	16 ♂	Proximally in L. tibia	None	Curettage + bone grafting	Consistent with non-osteogenic fibroma of bone
5	14 ♀	Proximally in R. tibia	None	Curettage + bone grafting	Non-osteogenic fibroma of bone
6	12 ♂	Distally in L. fibula	None	None	
7	13 ♂	Proximally in L. tibia	Pain in region of left knee	None	
8	19 ♂	Proximally in L. fibula	Uncertain pain in both knees	None	
9	15 ♂	Distally in R. tibia	None	None	
10	10 ♀	Proximally in L. tibia	Pain, discomfort in region of left knee	None	

sclerotic marginal zone increasing in thickness with the age of the lesion (Ponseti & Friedman 1949, Cunningham & Ackerman 1956).

The pathological appearances of a non-osteogenic fibroma of bone are as follows: one or several, more or less separated cavities filled with a "dry", yellowish-brown, brown, or grey tissue. The overlying cortex may show patchy erosion. No periosteal thickening. The demar-

Case reports .

Follow-up period (months)	Status at completion of treatment or last follow-up examination
28	Clinically symptom-free. X-rays: Small residual defect, otherwise the appearances had returned to normal.
17	Clinically symptom-free. X-rays: Small residual translucencies in the peripheral area, otherwise normalization.
12	Clinically symptom-free. X-rays: Complete normalization of the skeletal structure. Slightly angulation after fracture.
13	Clinically symptom-free. X-rays: Mild postoperative changes. No residual defect.
6	Clincally symptom-free. X-rays: Mild postoperative changes. No residual defect.
19	Clinically symptom-free. X-rays: Definite reduction of the defect. Distance from epiphyseal line increased by 1.2 cm.
10	No subjective complaints. Mild atrophy of the quadriceps. X-rays: Defect a little smaller. Distance from epiphyseal line increased by 0.5 cm.
36	Still mild pain in both knees. X-rays: Considerable reduction of the defect.
30	Clinically symptom-free. X-rays: Defect has disappeared.
48	Mild pain in the region of the left knee. X-rays: The first defect has become almost obliterated. The second defect has increased in size. Distance of both defects from the epiphyseal line increased.

cation from the normal cancellous bone is smooth and sclerotic. The microscopic picture shows the following characteristics: (1) Areas of firm connective tissue and a sparse intercellular substance. Commonly deposits of haemosiderin in the connective tissue. (2) Giant cells having from 3 to 10 nuclei. (3) Large and small, lipoid-filled macrophages (foam cells) in a loose connective tissue. (4) Complete absence of

bone formation in the lesion itself (except in cases of fracture through the lesion).

It is well known (Hatcher 1945, Robbins 1962) that the histological appearances vary somewhat from one area to another within the same lesion. Furthermore, it is assumed (Hatcher 1945) that the quantity of xanthomatous tissue increases with the age of the lesion. Lastly, it may be mentioned that the differential diagnosis from giant-cell tumour and fibrous dysplasia is difficult (Robbins 1962).

Lacking a classification and a definite knowledge of the aetiology, it is difficult to set up clear lines for treating non-osteogenic fibroma of bone. Jaffe & Lichtenstein, who interpreted the condition as a neoplasm, found indication for surgery in all cases. Later, the tendency to spontaneous healing has become known, and accordingly the therapeutic attitude has changed to a more conservative one. Today it seems to be agreed that only some of the cases require operation, and the following operative indications have been set up (Compere & Coleman 1957): defects which give rise to symptoms, which are unusually large, and which have caused pathological fractures. To this may be added: defects in which the X-ray diagnosis is in doubt and in which a biopsy is needed. All authors agree that curettage of the defect, with or without bone grafting, is adequate treatment. X-ray therapy is ineffective (Devlin et al. 1955).

PRESENT MATERIAL

The present material comprises 10 cases collected over a period of 10 years. Nine of these patients were treated in the Lillehammer County Hospital during the period January 1964 to December 1968. One patient (Case 1) had been treated in another hospital in 1958. The clinical data are listed in Table 1. Two cases will be reported briefly:

Case 4: A boy, aged 16, whose history was negative for major illnesses or complaints from the left knee or lower leg. When playing football on 8 September 1968 he sustained a severe trauma to his left knee. A few days later he was admitted with symptoms of rupture of the internal lateral ligament of the left knee. X-rays (Figure 2) revealed no skeletal damage, but a multilocular defect, 4 × 2 cm, medially in the proximal tibial metaphysis. The lesion was interpreted as non-osteogenic fibroma of bone. On 18 September 1968 suturing of a totally ruptured internal lateral ligament was done, and at the same time curettage and bone grafting of the defect in the tibial metaphysis. Histological examination (Dr. Reidar Eker, prep. No. 18710/68) compared with the X-ray appearances fitted the diagnosis of non-osteogenic fibroma of bone. When last seen, 13 months after

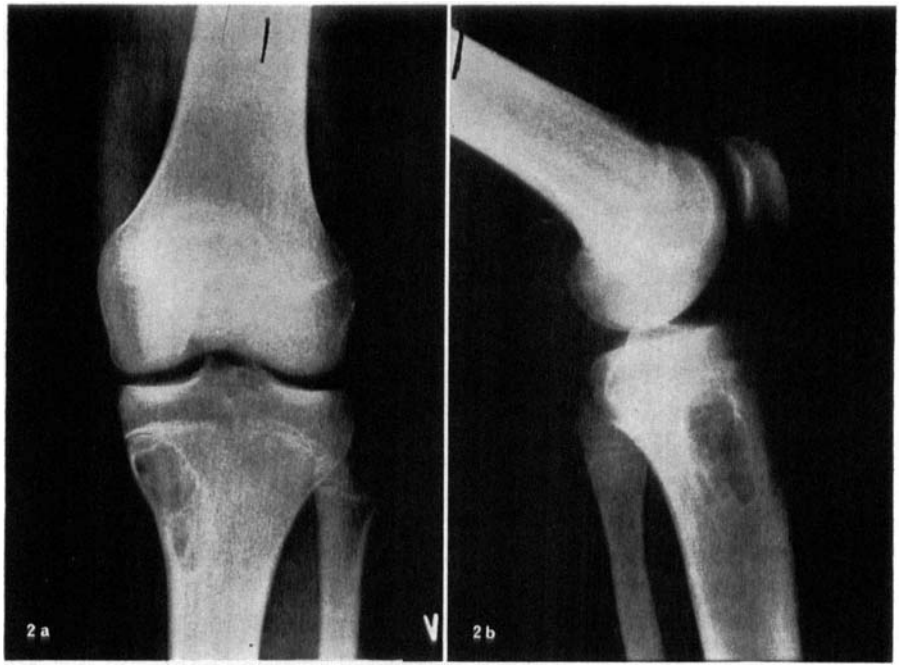
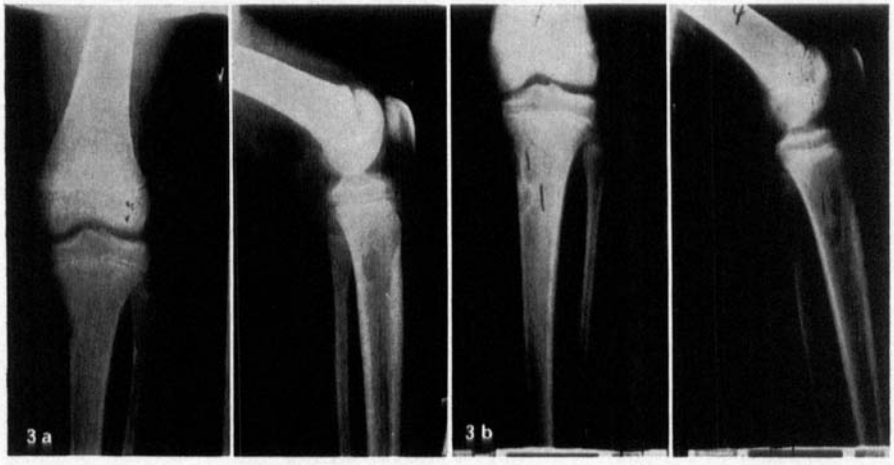


Figure 2 a and b. X-ray of left knee joint 9 days after acute injury
Note that the defect does not involve the epiphyseal line.

the operation, the patient had no clinical symptoms; X-rays showed postoperative changes, but no residual defect.

Case 10: A girl, aged 10 years, who had been treated 3 years previously by wire traction through the proximal end of the tibia for left-sided fracture of the femur. From the summer of 1964 she had noticed mild pain in the upper part of the left lower leg. Owing to these complaints, X-ray examination was done in October 1964. It revealed a well-defined defect, 2 × 1.2 cm, in the proximal tibial metaphysis (Figure 3 a). The defect was interpreted as a non-osteogenic fibroma of bone and observed at intervals without a biopsy. After several intervening follow-up X-rays had shown mainly stationary appearances, an X-ray in April 1966 revealed a new defect proximal to the first one which had "migrated", in the course of the follow-up period, 1.2 cm from the epiphyseal line (Figure 3 b). In October 1968 the first defect had become sclerotic and had almost disappeared, and its distance from the epiphyseal line had increased by another 1.5 cm. Defect No. 2 had increased considerably in size since its detection 30 months previously. At this examination a translucency was also found in the lateral femoral condyle, compatible with osteochondritis dissecans (Figure 3 c). During the period October 1964 to April 1968 the patient had no complaints of the left lower leg or knee. From the summer of 1968 she again developed mild pain in the region of the



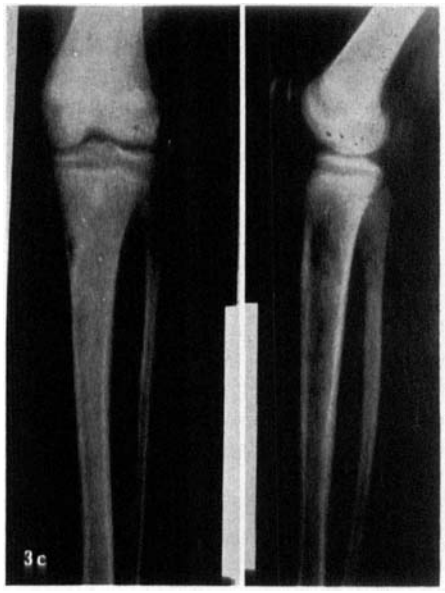


Figure 3 a. X-ray of left knee and lower leg in October 1964 showing an oval defect surrounded by sclerosis. Figure 3 b. A new X-ray 18 months later showing a smaller defect proximal to the original defect which has decreased somewhat in size. Figure 3 c. X-ray from October 1968 showing that the primary defect has almost disappeared, whereas the latter has increased in size. At the same time, osteochondritis dissecans was found in the lateral femoral condyle.

left knee, presumably due to osteochrondritis dissecans, since now the pain was localized in the knee joint.

COMMENTS

All the patients were between 9 and 19 years of age at the time of diagnosis, average 13.9 years. Three were females and seven males. All the defects were localized in the lower limbs, an equal number on the right and left. Seven defects were in the tibia, two in the fibula,

and one in the femur. Seven of the ten patients were symptom-free, while two complained of mild pain in the region. One patient presented with a pathological fracture. In five cases indication was found for operative treatment owing to the size of the defect. Neither of the two patients with pain had operative treatment because their symptoms subsided within a short time. Among the five patients who were followed for a long time by X-rays the defect subsided in one, diminished considerably in two, and slightly in one. In the last patient of this group the primarily detected lesion has almost disappeared, while a second one has increased in size. The last four patients are still being followed.

DISCUSSION

The cortical fibrous defects commonly seen in the metaphysis of children are not definitely identical with non-osteogenic fibroma of bone. However, some authors (Hatcher 1945, Compere & Coleman 1957) have found it unnatural to differentiate between the two types, because the histological appearances are similar, both types of defect show a marked tendency for spontaneous healing, and both conditions occur in the same age group. It may be objected to this view that histological examination of fibrous lesions of bone affords an unreliable basis of classification, because the microscopic appearances of these defects appear to vary a good deal, both according to the age of the lesion as well as area by area within the same preparation, and because histological assessment of benign skeletal lesions is on the whole difficult (Robbins 1962). It may be objected further that the fibrous defects described by Sontag & Pyle and by Caffey are of a typical cortical localization, have little tendency for perifocal sclerosis, and remain at a constant distance from the epiphyseal line, while non-osteogenic fibroma of bone is mainly of a subcortical localization, shows distinct perifocal sclerosis, and migrates away from the epiphyseal line as long as the line remains open. Another difference between the two types of defect is their incidence. While cortical defects occur in more than 50 per cent of all healthy boys and in almost 25 per cent of all healthy girls at some time or other between the ages of 2 and 18 years, non-osteogenic fibroma of bone appears to be an uncommon condition although there has been no direct study of its incidence. Pending further knowledge concerning the aetiology and pathogenesis of the fibrous defects, it seems natural to classify

them into two types and to name these two types cortical fibrous metaphyseal defects and non-osteogenic fibroma of bone respectively.

It is generally assumed that non-osteogenic fibroma of bone is a benign condition. However, there has been a report of a case of osteogenic sarcoma arising in a non-osteogenic fibroma of bone (Hastrup & Skov Jensen 1965). This report is interesting, but several factors indicate that the original lesion had been misdiagnosed. It was localized in the epiphyseal area, it was larger than a non-osteogenic fibroma of bone usually is, and the X-ray appearance of the original defect was not typical of this last-mentioned condition. The histological appearances—apart from showing malignant tumour tissue, areas of fibrillar connective tissue, giant cells, and foam cells—do not rule out that the primary lesion may have been a giant-cell tumour or fibrous dysplasia which would fit better in with the patient's age, the site of the lesion, and the X-ray findings.

The present material seems to confirm that non-osteogenic fibroma of bone is more common in boys than in girls and that practically all patients are in the age range 2–20 years at the time of diagnosis. The fact that the condition was silent in 7 out of 10 cases is in conformity with previous findings (Maudsley 1956, Compere & Coleman 1957). Lastly, the frequent localization of the defects in the tibia fits well with Compere & Coleman's material in which 14 out of 20 defects were in this bone. Case 10 exhibited several interesting features: two defects occurred at different times in a site where traction through the bone had been applied 3 years previously. Considerable enlargement of the second defect occurred during the follow-up period. Lastly, the patient had osteochondritis dissecans of the ipsilateral femoral condyle. In several of the patients who had no surgical treatment, the distance of the defect from the epiphyseal line increased in the course of a few years, and a number of these defects decreased considerably in size.

CONCLUSION

(1) Non-osteogenic fibroma of bone should be classified apart from cortical fibrous defects. (2) Lesions which give rise to symptoms and large lesions should be treated by curettage and bone grafting. (3) Smaller lesions which may be definitely diagnosed by X-rays may be observed at intervals.

SUMMARY

Non-ostegenic fibroma of bone is of unknown aetiology, but recent authors assume that it is due to disturbed ossification or necrosis left by infarction. The relationship between non-ostegenic fibroma of bone and the far more common cortical fibrous defects is in doubt; and pending further knowledge concerning the fibrous metaphyseal defects, the two types should be kept separate.

Subjective symptoms are not outstanding, and many lesions are discovered incidentally in an X-ray examination done because of trauma. The X-ray diagnosis is reliable in non-ostegenic fibroma of bone. On the other hand, it may be difficult to distinguish the appearances histologically from other benign skeletal diseases, in particular from giant-cell tumour and fibrous dysplasia.

A material of 10 patients is presented. Five were treated by curettage of the defect and were confirmed histologically. In the other patients the diagnosis was based exclusively upon the X-ray findings. One patient of this latter group developed two defects in an area where wire traction had been applied three years before the diagnosis; this patient had, in addition, osteochrondritis dissecans of the ipsilateral femoral condyle.

The indications for surgical treatment of non-osteogenic fibroma of bone are set up.

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