

LOCALIZED OSTEITIS DEFORMANS

By

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Among the many cases of osteomyelitis seen by the orthopaedic unit, there is a type of chronic osteomyelitis which has been of particular interest to us. There was no cavity or sequestra formation. The radiographic appearance and the visible deformation of the tibia led us to diagnose a very rare lesion described by *Lièvre* (1936) under the name of "Remaniement pagetoïde localisé d'origine traumatique".

It is a type of osteitis deformans, but remains localized and appears long after at the site of trauma. There follows a summary of two cases seen by us: the one involving the tibia, the other the fibula.

CASE REPORTS

Case 1. Miss V. L., aged 19, admitted to Lovanium clinic on March 19, 1959.

History. When three years old, the patient sustained mild trauma to the left leg which rapidly became a dirty wound with generalized signs of toxemia. She was hospitalized and curettage of the tibia was carried out; skin grafting was later necessary to close the wound which remained indolent. From the age of three to the time of admission, the formation of purulent ulcers frequently recurred interrupting the rare periods of apparent healing.

On admission. The patient presented with a painful swelling of the left leg which impeded walking. The leg was curved forwards. At the level of the anterior border of the tibia, the skin was stretched and atrophic with areas of hyperpigmentation.

At the levels of the middle and lower thirds of the tibia were two ulcers, each of approximately 4 cm. in diameter overlying the anterior border of the tibia.

The skin was warm and showed increased sweating; examination of the leg provoked excessive sweating of the lower leg within a few seconds. There was no joint involvement.

Investigations. On X-ray examination, comparison between the two tibias showed the following differences: a diffuse thickening of the whole diaphysis of the left tibia which was 1 cm. longer and bent forwards; the fibula was normal. (Figs. 1 and 2). In its middle third, the left tibia was twice as thick as the right one and it was difficult to distinguish between the cortex and the medulla, particularly at the anterior border. The structure of the bone was lamellar with an irregular criss-

Fig. 1.*Fig. 2.**Fig. 2 a.**Fig. 3.*

Fig. 1. The thickening of the left tibia is very important. Disappearance of the limit between cortex and medulla. Pseudo-cystic aspect of the lower third.

Fig. 2. Lateral view. The tibia is bent forwards and the structure of the anterior border is lamellar.

Fig. 2 a. Enlargement of one area of the Fig. 2.

Fig. 3. Postoperative aspect; hollowed out tibia.

cross pattern and minute cavities. (Fig. 2 a). The density of the bone was very unequal: opaque and transparent areas were irregularly disposed: there were no visible sequestra. The X-ray appearance of the skull, lumbar spine and the pelvis were normal. Routine blood and urine examination showed no abnormalities.

- E.S.R. (Westergren): 41 mm. in one hour.
- Serum phosphorus: 4.6 mg. %.
- Serum calcium: 10 mg. %.
- Serum acid phosphatase = 0.14 Bodansky units.

- Serum alkaline phosphatase = 3 Bodansky units.
- Wassermann: neg.—Widal: neg.
- Urine calcium (Sulkovitch) increased +++.

Discussion of treatment. The following aspects of this problem were considered:

- 1° The covering of the ulcers with skin.
- 2° The trophic disorders of the skin.
- 3° The deformation of the tibia.
- 4° The elimination of pain.
- 5° The determination of the nature of the lesion (bone biopsy).

To realize these five aims, we decided on the following operation: resection of the anterior border of the tibia with hollowing out of the bone on the anterior medial two-thirds, lateral mobilization of skin, suturing it over the thinned tibia.

Treatment. By a longitudinal, elliptical incision, the atrophic skin was excised, the medial surface of the tibia on two thirds of its length was removed; the anterior border of this bone was rectified. The bone consistency was very softened: the cortex was soft and the medulla was filled with cancellous bone, abundantly vascularized. A posterior lateral counter-incision was made to cover the bony surface with healthy skin. Later skin grafting was carried out to cover the defect of the counter-incision and a small area of necrosis.

Follow-up. The postoperative X-ray showed the limits of the excavation: on the lateral view, the tibia seems to be well reestablished. (Fig. 3). The posterior cortex remains and appears strong enough whereas an important part of the fibrillar bone is removed. Seen recently, the patient was satisfied with the result: pain disappeared and walking was easy. The scar was good but the skin in this area was thin, too pigmented and partially adherent to the deep tissues. The general condition was quite excellent and control blood and urine examinations gave similar results: the excessive calcium elimination in the urine persisted.

Case 2. Mrs. P. N., 45 years of age.

History. When she was about twenty years old, this patient sustained an injury just above the lateral malleolus which became ulcerated. She was hospitalized elsewhere for one month before healing occurred. Twentyfive years later, in 1958, a new ulcer appeared at the same site which persisted until admission to our hospital on April 27, 1959.

On admission. The patient presented with a swelling of the ankle and of the lower third of the left leg. Above the lateral malleolus, an old ulcer of 2 cm. of diameter and 0.5 cm. deep was situated over the fibula. This whole area was warmer and showed the same local symptom of hyperhydrosis as the first case. Walking was painful. The fibula was thickened and tender in its lower third. There was no limitation of movement at the ankle joint.

Investigations. On X-ray examination, the left fibula was normal in its upper third: lower it progressively thickened until its diameter was equal to that of the tibia. (Figs. 4 and 5). In its lower third, the cortex could not be distinguished from the medulla: the outline of the bone was very irregular, hazy in some places, and the degree of calcification was unequal. There were no sequestra visible. The joint appeared normal but the lateral cortex of the tibia appeared to be slightly involved.

*Fig. 4.**Fig. 5.**Fig. 6.*

Fig. 4. Progressive thickening of the fibula from top to bottom.

Fig. 5. The lower third of the fibula has the same width as the tibia. The outline of the bone is irregular and hazy.

Fig. 6. Postoperative radiography.

The X-ray appearances of pelvis and skull were normal; the lumbar spine showed some lesions of arthrosis.

Routine blood and urine examinations gave normal results.

- E.S.R. (Westergren) = 40 mm. in one hour.
- Serum phosphorus = 3.4 mg.‰.
- Serum calcium = 9 mg.‰.
- Serum acid phosphatase = 0.18 Bodansky units.
- Serum alkaline phosphatase = 3.9 Bodansky units.
- Wassermann = neg.—Widal = neg.
- Urine calcium (Sulkovitch) increased ++.

Treatment. The therapeutic problem was similar to that of the first case: to relieve the pain and the ulcer, to reduce the deformation of the lower leg and to study the histology of the bony lesion. On May 9, 1959, the ulcer and the lower two-thirds of the fibula were excised under tourniquet: a part of the lateral malleolus was left to avoid distortion of the ankle joint. (Fig. 6).

The wound healed rapidly and the patient walked normally, without pain. Seen again in May 1960, ankle joint motion had remained free and painless.

HISTOLOGY EXAMINATIONS

Professor *J. Vincent* of Lovanium has examined the bone biopsies of both patients. The micro-radiographic aspect shows an irregular Haver-

sian architecture which resembles Osteitis Deformans of Paget, but which is nowhere characteristic. Generally, there is a distinct lamellar structure. The walls of the Haversian systems are frequently crossed by small vascular canals. The amount of calcium in one haversian system and from one to another is variable just as in a normal bone. (Fig. 7).

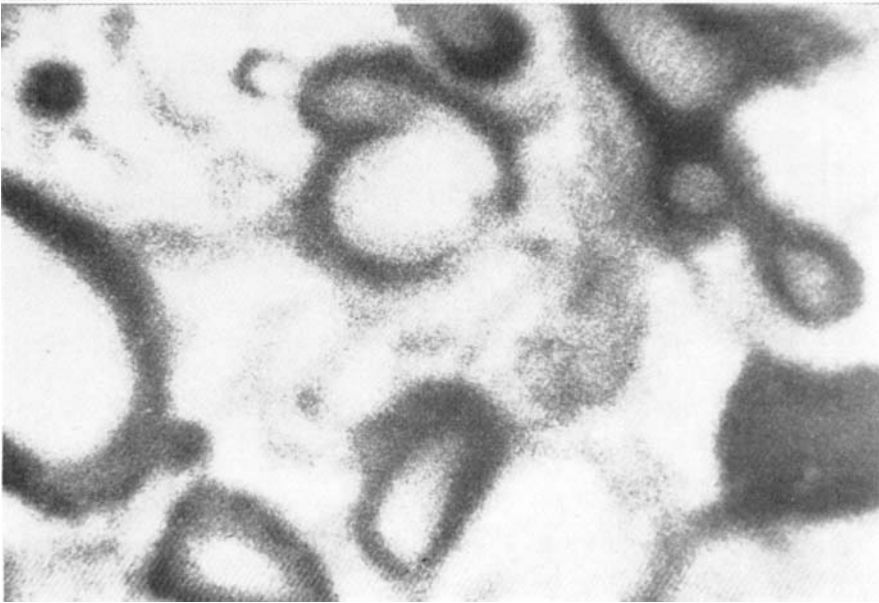
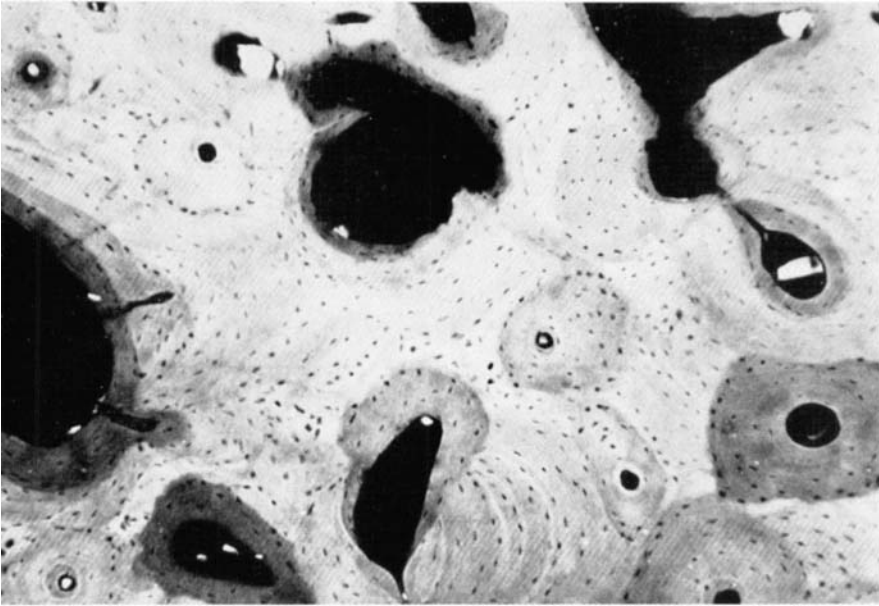
Mc Manus method applied after decalcification of the fragments shows that coloration with PAS varies inversely in intensity with cal-



Fig. 7.

Correlation between the microradiography (7-A) and the histological picture of the same slide after decalcification and coloration with PAS (7-B). The structure of the tissue is disturbed. Two arrows indicate vascular canals in the wall of Haversian systems. The essential bone substance is the more colorated by the method of Mc Manus that it was less calcified. Enlargement 66 $\times$.

A



B

Fig. 8.

Correlation between the microradiography (8-A) and the autoradiography made in vitro with S^{35} (8-B). Lamellar disposition. The radioactivity is in the less calcified tissue. Enlargement: 75 $\times$.

cification. Some preparations were dipped in vitro in a solution of $\text{Na}_2\text{S}^{35}\text{O}_4$. Recorded on a Dentus Rapid (Gevaert) film, the autoradiography was compared with the microradiographic picture: it showed that the affinity for the S^{35} was limited to areas incompletely mineralized. (Fig. 8).

The day before the operation, the second patient received intravenously Aureomycin (25 mg. per kilo) in order to observe the bone fluorescence. The study of these sections in the extreme blue light brings out fluorescent rings around the largest of the haversian canals. These rings indicate in dogs the sudden and massive deposit of mineral salts. (*Ghosez*).

In conclusion, these special investigations show no abnormalities in the constitution of the bone substance itself but they illustrate the irregular pattern of the haversian system similar to that seen in Osteitis Deformans.

The pictures of the two cases are similar.

DISCUSSION

Whereas the radiological appearance and the deformation of the tibia are in accord with the osteitis deformans of Paget, the histological aspect is a little different and leads us to consider a lesion described in 1936 by *Lièvre* under the name of "Remaniement pagetoïde localisé d'origine traumatique".

In 1936, *Lièvre* described the clinical and radiological signs, but it was only in 1957 that *E. Rohner* published the results of his histological investigations: he found a fundamental difference in the bone structure of the two lesions. In the osteitis deformans, the structure of the bone is formed by many small irregular fragments, all connected together by ramified fibres, giving the pathognomonic aspect of "Mosaic Pattern".

In contrast, in the localized "Remaniement pagetoïde" the mosaic pattern is rather rare and not as typical: the bone is constituted by lamellae directed longitudinally: the connecting fibres are not as numerous.

The particular aspects of this syndrome are the following: initial trauma, localization, free interval, sex and age of the affected patients, clinical and radiologic signs. We intend to develop it and compare with the history of the two preceding cases.

— The initial trauma may be mild or violent: it causes at least a bony contusion with a sub-periosteal hematoma. Sometimes it is strong enough

to cause a fracture as in the three cases of *Rohner*. In our two cases, the trauma was mild but was immediately complicated by serious infection. This inflammatory aspect, the long suppuration without elimination of sequestra is quite atypical and constitutes a particularity of our two African cases.

— The most frequent localization is the tibia but others have also been described: fémur, skull . . . and simultaneous involvement of the tibia and the fibula. The superficial situation of these two bones exposes it to trauma and would explain their relative frequency.

— The free interval usually lasts several years. It is quite characteristic for the localization on the fibula where for twentyfive years, the patient had no complaints. For the localization on the tibial diaphysis, the interval was also of many years but this interval was not free: frequent ulcers occurred during these sixteen years.

— Generally, women over the age of forty are affected. Our first case with the age of three for the initial trauma and nineteen for the constituted lesion with bending of the tibia does not accord with this picture.

— The clinical signs are convincing: bone pain, important hypertrophy of the involved bones, bending of the tibia and local vasomotor disorders: skin hyperthermia, hyperhydrosis and abnormal pigmentation.

— The radiological signs are similar to those seen in osteitis deformans: hyperplasia of the bone in width, increased length, changes in the structure of the bone with disappearance of the limit between cortex and medulla, unequal degree of calcification . . .

What is the pathogenesis of this lesion? What is the underlying mechanism of its formation? Generally, it is believed that the initial trauma causes vasomotor disorders in the involved bone. *Rohner* points out this lesion could be a type of Sudeck's atrophy. This hypothesis, however, can be discounted by the poor results given by sympathetic infiltrations and sympathectomies.

Treatment is generally disappointing: sympathectomies, radiotherapy give no success. Local treatment may only be attempted, for instance corrective osteotomy.

Before the inflammatory aspect of our two cases, we did not hesitate to excise the diseased skin and to remove an important part of the affected bone. Our surgical treatment stopped the infected ulcers and the pain. The long-term prognosis remains, however, very reserved because we did not stop the vasomotor disorders and the bone altering as the urine calcium elimination tests showed.

SUMMARY

Two further cases, the first in Africa, of "Remaniement pagetoïde localisé" are described. The number of cases now published is about twenty. Modern histological methods confirm this as a well-defined disease entity.

Treatment by wide excision of bone gave a symptomatic cure in both these cases.

RESUME

Deux nouveaux cas, les premiers en Afrique, de "remaniement pagetoïde localisé" sont décrits. Le nombre des cas publiés jusqu'ici s'élève à environ 20. Les méthodes histologiques modernes confirment qu'il s'agit là d'une bien définie.

Le traitement par large excision osseuse a amené la guérison dans ces deux cas.

ZUSAMMENFASSUNG

Zwei weitere Fälle, der erste in Afrika, von „Remaniement Pagetoide Localisé“ werden beschrieben. Die Zahl der bisher veröffentlichten Fälle ist ungefähr zwanzig. Moderne histologische Methoden bestätigen die Erkrankung als eine wohlabgegrenzte Einheit.

Die Behandlung mit weitgreifender Excision ergab eine symptomatische Heilung in beiden Fällen.

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