FROM THE ORTHOPAEDIC CLINIC, HÄRNÖSAND
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MELORHEOSTOSIS

Report of a case

By

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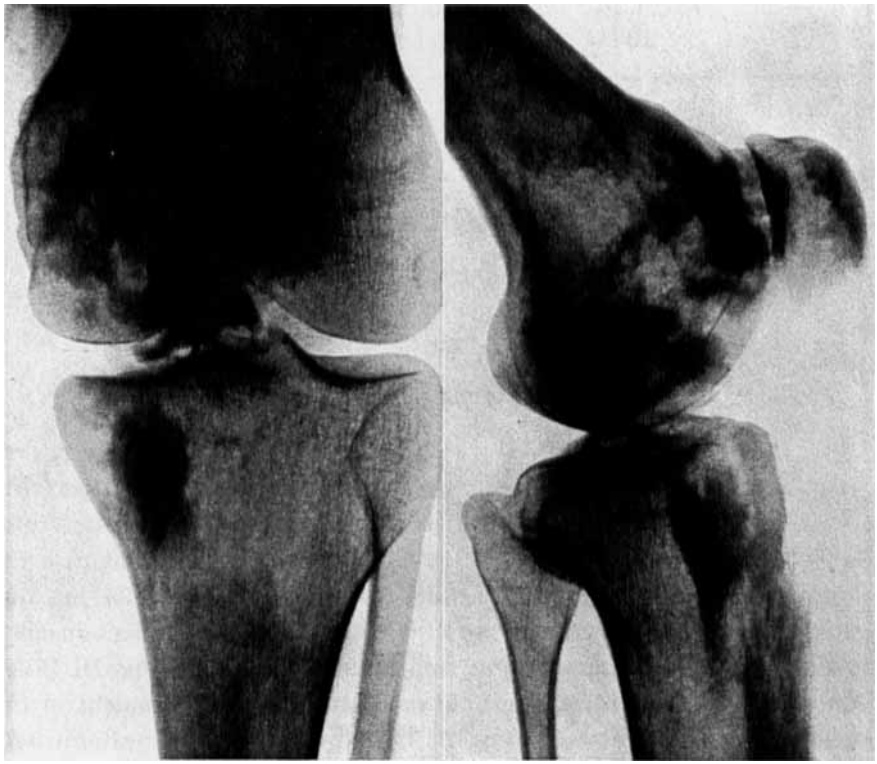
A man aged 49 is the subject, a woodsman and manual worker, who ever since his childhood has been troubled by a certain stiffness and feeling of tiredness in his left knee and his left foot. He has also experienced a longish period with a difficult eczema. After 1941 he was examined on several occasions at the orthopeadic department in Härnösand and he complained about aching and stiffness in his left knee joint and foot. At several examinations limitation of movement in the knee joint and some thickening of the capsule could be established. On 10.10.57 a follow-up was instituted and it proved that mobility in the left knee joint had considerably lessened and at the same time the pain had increased. It was difficult for him to carry out his work. The range of movement in the knee was 150°–90°. X-ray examination showed significant progress of the strata typical of melorheostosis. These had, as it were, slipped down in the joint cavity so that exostosis formation was present (see fig. 1). On the inner aspect of the foot and lower leg a projection was palpated and there was moderate pain on pro- and supination movement in the tarsus. Radiology showed typical changes here too (see fig. 2). As regards the increasing trouble from the left knee operation was proposed. This took place 18.10.57. Medial incision. The capsule was considerably thickened, greyish white and hardened with strong fibrosis. At the back of the medial femoral condyle a large, mushroom-like exostosis was found and a similar, smaller one in the anterior part of the condyle, plainly projecting from the changes here too (see fig. 2). As regards the increasing trouble from away, after which the knee could be extended; but 10° extension defect remained in spite of this. At the follow-up the patient reported considerable improvement and he was capable of returning to his work.



A *Fig. 1.* B

Here a case of melorheostosis is involved, a very rare complaint, which was described for the first time by *Leri & Joanny* 1922. In the typical cases the changes are localized to one of the extremities. The contour of the affected leg is sooner or later distorted. The name was suggested by *Leri & Joanny*. It was their opinion that the change resembled half liquid wax, hence the name. *Putti* published in 1927 two cases, both localized to the lower extremities. He gave the change the name of osteosi eburnizzante monomelica. Further cases were published by *Ljunghagen* 1930, *Wakeley* 1931, *Boggon* 1938, *Franklin & Matheson* 1942, *Fairbank* 1948, *Thompson, Allen, Andrews & Gillwald* 1951, and *Campbell* 1955.

The etiology is unknown. *Putti* suggested that the cause is ischemia secondary to local changes in the sympathetic nerve system.

The disease is more common in men but also occurs in women and cases have been described from the age of 5 to 50; it is probable, however, that the condition arises in childhood and possibly even prior to birth (*Ljunghagen* 1930).

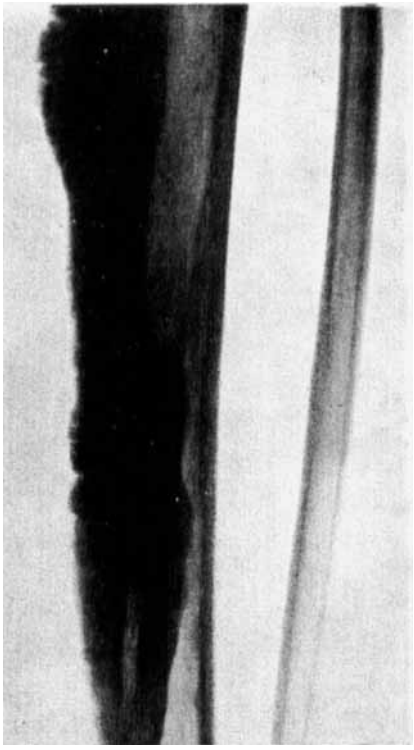


Fig. 2.

Localization: the upper extremities are less affected than the lower and it is typical that the changes are mostly localized to one extremity. Fairbanks states that he found 7 cases in the literature where the localization occurred in more than one extremity.

The pain is the most characteristic feature. It is aching and continuous, often quite severe. *The limitation* of movement was plain in half of the cases (*Fairbank* 1948). The cause of this limitation of movement is, as in the case described, often exostosis formation but in addition the affection of the joint capsules. A change in the configuration of the extremities occurs often with oeedema, but deformities of the type genu valgum or varum seldom arise. Scleroderma with fibrosis and thickening of muscles and other tissues have been described in a number of cases (*Thompson, Allen, Andrews & Gillwald* 1951). The present case also experienced over a period a very troublesome eczema, which may well have been just such a change in the soft tissue.

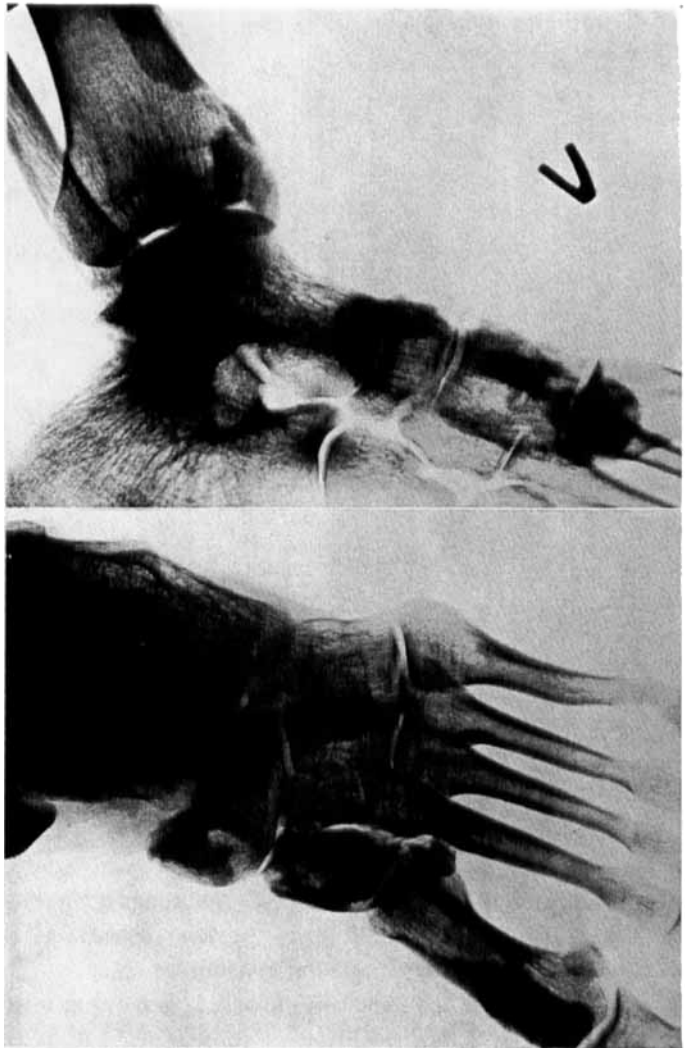


Fig. 3.

Radiological changes: in typical cases sclerotic, featureless strata of the type "marblebones" are found. The previous description of liquid wax characterizes the radiological picture. One gains the impression that these strata follow the vessel and nerve paths. In the epiphyses and in the short bones, e.g. in the foot or hand, the strata alter their character and resemble spots, streaks or blurs. It is characteristic that

a certain progress occurs but it is very slow. Such was the condition in the case described.

The pathological picture is very specific and shows a strangely patterned picture with lamellae in irregular constructions. A mixture of mature and immature bone is found.

The diagnosis in typical cases is easy. As against general osteopetrosis the disease is distinguished by its localization usually to one extremity. In these cases therefore the whole skeleton should always be X-rayed. The differential diagnosis from osteopetrosis is normally easy. The latter disease is a general affection of the skeleton and is not bound to one extremity as melorheostosis often is.

SUMMARY

Melorheostosis is a rare disease which is mainly localized to one extremity of the skeleton, usually in the lower extremities. It is characterized by pain and limitation of movement which is often very troublesome in advanced cases. Presumably the disease begins prior to birth or in early childhood and progresses very slowly. In the case described above the changes were localized to the left knee joint where there gradually occurred a considerable limitation of movement, which could partly be remedied by chiselling away the protruding bone strata. The diagnosis is typical. The radiological picture has been compared to liquid drops of wax which, as it were, run down the long bones and are localized along the vessel and nerve paths. In the small bones the changes are of somewhat different character and resemble ink blots or are streaky.

RESUME

La mélorhéostose est une maladie rare localisée essentiellement dans les os des extrémités, habituellement des extrémités inférieures. Elle se caractérise par des douleurs et une limitation de la mobilite souvent considérable dans les cas avancés. Il est probable que la maladie apparaît déjà au cours de la vie fétale ou très tôt dans les années de l'enfance et qu'elle progresse très lentement. Dans le cas décrit ci-dessus, les modifications sont localisées au genou gauche, dans lequel il est peu à peu apparu une limitation notable de la mobilité qui peut cependant être partiellement supprimée par le grattage de la production osseuse anormale. Le diagnostic est typique. La radiographie montre

quelque chose qui ressemble à des gouttes de cire liquide qui paraissent couler dans le canal des os longs et sont localisées sur les vaisseaux et les filets nerveux. Sur les petits os, les modifications ont un caractère à peu près analogue et ressemblent à des pâtés d'encre ou à de petites taches.

ZUSAMMENFASSUNG

Die Melorheostose ist ein seltenes Leiden, das sich wesentlich im Skelett einer Extremitet, gewöhnlich in den unteren Extremiteten, lokalisiert. Sie ist durch Schmerzen und Bewegungseinschränkung charakterisiert, die in fortgeschrittenen Fällen oft sehr beschwerlich sind. Wahrscheinlich tritt die Anlage zur Erkrankung bereits beim Fötus oder im frühzeitigen Kindesalter auf und schreitet sehr langsam fort. In dem oben beschriebenen Falle sind die Veränderungen am linken Kniegelenk lokalisiert, wo nach und nach eine bedeutende Bewegungseinschränkung auftrat, welche doch teilweise mittels Abmeislung der hervorstehenden Knochenauflagerungen behoben werden kann. Die Diagnose ist charakteristisch. Das röntgenologische Bild wurde mit Wachstropfen verglichen, die sozusagen entlang den langen Röhrenknochen fliessen und an den Gefäss- und Nervensträngen lokalisiert sind. An den kleinen Knochen zeigen die Veränderungen ein etwas verschiedenes Aussehen und gleichen Tintenklecksen oder kleine Striche.

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