MELORHEOSTOSIS OF THE UPPER LIMB

Report of a Case

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Melorheostosis is an unusual type of bone petrosis which typically affects the bones, or parts of bones, of one limb. The name was given to the condition by Léri and Joanny (1922), who recorded the first case. Fairbank (1951) considered that, although over forty cases had been recorded in world literature, only four authentic cases had been presented in the United Kingdom. These were the cases of Wakeley (1931), in which both lower limbs were affected; of Boggon (1938), in which the right lower limb was involved; of Franklin and Matheson (1942), in which an extensive petrosis occurred in the skull, spine, pelvis, and the bones of the arm and leg, all on the right side; and of Le Vay (1946), in which the site of the condition was the left lower limb. Three further cases have since been recorded in the British literature. The case of Thompson, Allen, Andrews, and Gillwald (1951) involved the right lower limb, that of Trevethick (1953) the left lower limb, and that of Elkeles (1954) all four limbs. Of these seven patients only two, those of Franklin and Matheson and of Elkeles, displayed an upper limb involvement. The following case of melorheostosis in the right arm and shoulder girdle was considered worthy of record because of the site and rarity of the condition.

CASE REPORT

A coal-miner aged twenty-six years complained of pain and stiffness in the right thumb for one year. He admitted to transient pain and stiffness in the right shoulder for eight years. He had no other complaints.

Examination revealed a thickened right thumb with marked limitation of movement of the metacarpo-phalangeal joint and slight limitation of the interphalangeal joint. There were no other notable abnormalities of the limb.

Radiographic investigation demonstrated the typical “candle grease flow” of melorheostosis. The whole skeleton was radiographed and the petrosis was found to affect only the right upper limb (Fig. 1). The density was evident in the coracoid process of the scapula, the upper half of the humerus, the lower end of the radius, the carpus on the radial side, and all the bones of the thumb. Bone formation was present in the soft tissues adjacent to the clavicle. The bony outlines of the phalanges of the thumb were irregular and the radius was thickened and slightly distorted (Figs. 2 to 4).

After one year a slight but appreciable increase in the petrosis was noted in the coracoid process of the scapula and the phalanges of the thumb. This evidence supports Léri and Joanny’s findings of progression in their original case of affection of the upper limb.
Fig. 2
Radiograph of the shoulder showing new bone formation in the region of the clavicle and the petrosis in the coracoid process and the humerus.

Fig. 3
Radiograph of the forearm and hand showing the “flow” of the petrosis and the thickening of the radius.

Fig. 4
Radiograph of the hand showing the degree of involvement of the thumb and the distortion of the phalanges.
COMMENT

The features distinguishing melorheostosis from generalised osteopetrosis were described by Fairbank (1951): "1) The changes, in typical cases, are confined to the bones of one limb; 2) the outline of the affected bone is, sooner or later, distorted; 3) there is often pain, occasionally severe, sometimes unbearable; 4) there is limitation of movement in the joints formed by the affected bones." The present case displays these features and also illustrates the way the petrosis may skip a joint, here the elbow; the formation of bone in the soft tissues, seen in the region of the clavicle; and the thickening and irregularity of a bone not markedly sclerosed. This last finding in the radius is comparable to the fibular thickening in the case of Le Vay, who suggested that it might be a pre-sclerotic effect.

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REFERENCES