Melorheostosis and the Sclerotomes: A Radiological Correlation


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Abstract. Melorheostosis is characterised radiologically by hyperostotic linear densities in bone. These densities have a peculiarly segmental distribution which does not correspond with the anatomical course of blood vessels or mixed nerve roots of the limbs. So far this distribution has lacked any valid explanation, although it has been suggested to be a developmental error as a result of an embryonic metameric disturbance. Inman and Saunders in 1944 described a sensory nerve supply to skeletal structures with 'sclerotomes' representing the zones of the skeleton supplied by individual spinal sensory nerves. Radiographs of 30 cases from the Radiological Museum of the Institute of Orthopaedics, London, have been reviewed and an attempt has been made to correlate the sclerosing lesions with the sclerotomes. The investigation was handicapped by paucity of films and clinical information, but in 19 cases the skeletal abnormalities were considered to correspond with a single sclerotome or part thereof. These studies were convincing when films of an affected hand or foot were available. In the remaining 11 cases multiple sclerotomes appeared to be involved and the clinical manifestations were correspondingly more severe. It is proposed that melorheostosis may be the late result of a segmental sensory nerve lesion, to account for its sclerotomal distribution. The association with linear scleroderma is discussed, since it has been suggested that these cutaneous lesions are related to the same nerve segment. Eight cases showed para-articular ossification of soft tissues which may be related to involvement of a corresponding myotome.

Key words: Bone, sclerosis - Melorheostosis - Ossification, ectopic - Sclerotome - Scleroderma, linear - Nerve, segmental sensory.

Melorheostosis is a rare mesodermal disorder, characterised typically by abnormalities of the skeleton and adjacent soft tissues. The entity was described originally by Leri and Joanny [6] in 1922. With the series now described the number of cases recorded in the literature approaches 200.

The clinical features vary widely. Chronic pain in an affected limb usually is a major complaint but, on occasion, the condition may be totally asymptomatic and be recognised only by incidental demonstration of pathognomonic radiological abnormalities of the skeleton. On the other hand, osseous excrescences and contractures of the soft tissues in advanced cases may cause orthopaedic complications sufficiently severe to require amputation. Various lesions of the skin have been reported in association with the disorder.

The disease usually is limited to a single limb, of which several bones may be affected. A consistent feature is the presence of clearly defined sclerotic densities in one segment of the bone or series of bones, having a linear pattern of distribution in the long axis of the limb. These densities are frequently peripheral with localised cortical thickening. The appearance of these linear hyperostoses has been likened to candle-wax flowing down the margin of the affected bone and extending often into the small bones of the hand or foot. Ectopic bone may be present in para-articular soft tissues. This 'peculiarly segmental distribution' does not correspond with the anatomical course of blood vessels or mixed nerve roots of the limbs [14] and so far has lacked any valid explanation. In 1927, however, Zimmer [20] suggested it to be a developmental error as a result of 'embryonic metameric disturbance'. This hypothesis was supported by Campbell et al. [1], in an extensive review of the subject. These authors observed that the distribution of the lesions in the bones of the limbs resembles that of paraxial hemimelia and suggested that the disorder is congenital, postulating 'that it is initiated early in embryonic life prior to formation of limb buds'.
Similar observation of a segmental pattern in the limbs was made by Inman and Saunders in 1944 [5] in relation to the radiation of deep referred pain from skeletal structures. They described such radiation of pain as being 'definitely segmental in character', yet unrelated to the distribution of any major nerve or blood vessel in the limb. From their experimental and clinical studies, Inman and Saunders derived the concept of a sensory nerve supply to skeletal structures related to spinal segments. The radiation of referred pain could be explained in terms of 'sclerotomes' or zones of the skeleton supplied by individual spinal sensory nerves. Their sclerotome maps (Figs. 1 and 2) exposed a segmental pattern of skeletal innervation, which is normally obscured by admixture of the segmental contributions within major nerve trunks. In effect, these workers unravelled the major nerves into their segmental components, and thereby revealed a pattern of skeletal innervation equivalent to dermatomes and myotomes.

Segmental distribution of pathology has been observed in a third clinical entity. A longitudinal axis of deformity in dysmelia—largely attributed to the toxic effects of thalidomide on the embryo during early pregnancy—was described by Henkel and Willert in 1969. This pattern, radiologically, was considered to be segmental [9, 10, 11]. Application of the sclerotome maps to radiographs of this group of reduction deformities has provided a rational explanation of their morphology [9]. Failure of formation of longitudinal segments of the appendicular skeleton can be interpreted as 'sclerotome subtraction' [11]. This failure implies some insult or injury to a segment or segments of the neural crest during embryogenesis (embryonic neuropathy) as the underlying mechanism of pathogenesis [7, 8, 10].