FROM AN ATLAS OF GENERAL AFFECTIONS OF THE SKELETON

14. OSTEOPATHIA STRIATA

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This very unusual affection is characterised by striation of the skeleton and particularly of the metaphyses of long bones. In 1924 Voorhoeve published three case reports, a father and two children, in which radiographs showed striation of the metaphyses with other minor changes in the bones which had not been described before. Shortly afterwards, this author published a report of the clinical findings in a boy aged twelve years with predominantly unilateral changes in which striation was a prominent feature (Fairbank 1925) and Dr Voorhoeve agreed that the case was similar to those he had described. Striation of the bones was also a striking feature in an unpublished case brought to my notice by Dr Grace Batten, but it differed in many respects from those mentioned above and will be considered separately, with a similar case published by Uehlinger (1941).

In 1935 the author suggested osteopathia striata as a title for this obscure condition. Two rather similar cases, a brother and sister, were published by Lindbom (1942) under the title of striated osteopoikilosis (Voorhoeve) but they are exceptional in many ways, being neither quite like Voorhoeve's cases nor like osteopoikilosis. Lindbom believed that disseminated dermato-fibrosis in his two cases reinforced the concept that the striation was related to osteopoikilosis. Hereditary and familial influences were evident only in Voorhoeve's original three cases. Sex—Three of the four cases were males. Age—Three of the cases were young, their ages being ten, twelve and fourteen years respectively: the fourth was the father of two of these children. Etiology—The cause is unknown: the author considers that it is the result of a congenital developmental error. Voorhoeve suggested that the striated bones he had described were related to dyschondroplasia and osteopoikilosis, the three disorders being variants of the same fundamental error, with the first forming a link between the other two, but this writer hesitates to accept such a view, particularly with regard to osteopoikilosis. Clinical features—There appear to be no symptoms definitely referable to the bone dysplasia. Vague recurrent joint pains, with or without a little swelling, were complained of in the hip and knee joints of one patient and in the shoulder joint of another. The author’s patient had suffered rheumatic fever affecting his heart, and in this unilateral case an abnormal gait and "awkwardness" had attracted the attention of the parents to the affected limb for some years and for this reason radiographic examination was made. The limb was longer and thinner than on the opposite side, but the upper limbs were of equal length. Otherwise there were no abnormal physical signs. Blood examination—Nothing of importance was discovered. Radiological examination—The outstanding feature is striation affecting in varying degree all bones with the exception of the skull and clavicles. In the long bones the striation affects principally the metaphyses: the dense lines run parallel to the long axis of the bones and can be traced for a considerable distance into the shafts. The thickness of the striations varies but most of them are fine and linear. Between the lines the bones may be porotic. Voorhoeve called attention to the appearance of both clear areas and dense spots in certain bones; and he pointed out that in some places one of the dense lines appeared to break through the cortex. The striation may or may not extend into the adjacent epiphysis. Sometimes the epiphyses are mottled with dense and clear spots. The clavicles are not affected. The hands are less affected than the rest of the skeleton. Striation is well seen in the tarsus and especially the calcaneus. The patella may be affected. In at least one of Voorhoeve's cases,
the boy of fourteen years, the ilia were striated with lines radiating towards the crests like
the columns of cartilage in dyschondroplasia. In our unilateral case there was a dense patch
in the ischium on the side on which the striation of the long bones was more obvious, and a
few irregular dense spots in the opposite ischium, these features being even more marked in
radiographs taken some years later. Usually the cortex of the bone is of normal thickness
and density, and there is no distortion. Voorhoeve, however, called attention to the presence
of exostoses in his cases; these were very small and were not found in the other case.

**Pathology**—So far, no pathological material has been available for investigation but it
seems probable that microscopic examination would reveal only normal bone of varying
density. Striation of the metaphyses, in our case at any rate, does not appear to have been
the result of any epiphyseal fault: early radiographs of the lower femoral and upper tibial
metaphyses showed that the most recently formed bone, to a depth of half an inch, was less
dense than that formed earlier; and that these clear bands were largely free from the dense
striation visible in the adjacent parts of the shafts; whereas radiographs taken four years
later showed no clear bands in these bones, and the striations extended through the metaphyses
to the epiphyseal lines. Thus, the dense lines had developed after the bone forming the clear
bands had been ossified. The region of the knee joint was the only place where juxta-epiphyseal
clear bands were visible in the early films.

**Diagnosis**—In dyschondroplasia, the bone between the columns of unossified cartilage often
gives rise to a striated appearance but this is never seen with such regularity in every major
metaphyses as in osteopathia striata. Moreover, in osteopathia striata the long bones of the
hand and foot are less affected than the major long bones; whereas in dyschondroplasia the
hands, and to a lesser extent the feet, are the chief seats of changes which could never be
described as striation. Reference has already been made to the fan-like arrangement of
striation which may be seen in the ilia in both these disorders.

The few dense spots seen in these cases are a little suggestive of those seen in
osteopoikilosis; but in typical cases of osteopoikilosis certain parts of the skeleton,
particularly the epiphyses, are closely peppered with dense spots. It is true that the spots
in osteopoikilosis may be prolonged in some places into streaks but these are much shorter
and much less regular than the dense lines seen in osteopathia striata.

Striation may be seen occasionally in one or two long bones in several other conditions but
it is never distributed widely throughout the skeleton as in this affection. In melorheostosis
dense streaking may be seen, but only to a very limited extent and only in one or two bones,
while the character and limited distribution of the other changes are usually typical. In
diffuse fibrosis of bone (polyostotic fibrous dysplasia), patches of increased density ending in
two or three broad streaks may be seen in the radiographs but only in one or two bones.

The later films of the knee joint on the more affected side in our case showed some
enlargement of the femoral and tibial shafts (enlargement of the fibula was noted in the
earlier films), and the radiographic picture had become a little suggestive of that seen in
diffuse fibrosis. The possibility that the affection in our case may not be identical with that
responsible for Voorhoeve's cases cannot be entirely discarded. Similar increase of density,
ending in streaks, may be seen in one or two long bones in neurofibromatosis; but this should
be recognised without difficulty by other features. We have seen striation in the region of
the knee joint in an atypical case of osteopetrosis. In Paget's disease an appearance worthy
of the name of striation is exceptional: but it may be quite marked in the os calcis at early
stages of the disease.

**GENERALISED HYPEROSTOSIS WITH PACHYDERMIA**

In 1941 Uehlinger published the report of a case of "hyperostosis generalisata mit
pachydermie" affecting chiefly the long bones, with coarse striation of a considerable part
of the skeleton. Dr Grace Batten's case, referred to above, appears to be identical except
that there was no pachydermia. Uehlinger referred to a number of other cases, some published before the discovery of X-rays, which he believed to be similar to the one he described. A familial tendency was apparent in many of them; and skin changes, affecting particularly the distal segments of the limbs, were present in some but not all. Other writers have linked these cases with pulmonary hypertrophic osteoarthropathy (for example, Freund 1938, despite the absence of changes in the chest and the marked differences in the radiographic appearances) but Uehlinger did not accept this view. Among the cases published with radiographs we have failed to find a single case that was strictly comparable with the patient reported by Uehlinger: in none was striation an outstanding feature. Uehlinger’s patient was a farm worker who died at the age of fifty-five years, the condition of the bones having been discovered incidentally at the age of forty-one years when a radiograph had been taken during the treatment of an infected toe. Difficulty in walking over rough ground, and undue fatigue, had been noticed for about one year. There was disproportionate length of the lower limbs; and both forearms and legs were noticeably stout, the skin being thick and leathery. The spine was very stiff, chest expansion was limited, and there was limitation of movement of several other joints. Later, the patient complained of constant pain in the bones, and in the abdomen, together with difficulty in micturition which was attributed to spinal compression. The contracture of the limbs increased and he became bedridden. There was bilateral cataract, and clubbing of the fingers, but no calcification in the kidneys or arteries. Little advance in the bone changes occurred during the last ten years of his life. After death, examination of the bones showed that the marrow was fatty and not fibrotic; the hyperostotic bone was mostly of the woven type. Dr Batten’s case, also a male, was above the average in height; his legs showed no obvious thickening and there was no dermatitis.

The cause of the condition seen in these two cases is unknown. In the second case there was a history of long-standing semi-starvation and the physicians considered that he had probably suffered from protein deficiency over a period of years.

The radiographic changes in these cases are of two kinds: 1) coarse striation of cancellous bone; and 2) thickening of the cortices of long bones. The striation is much coarser than that seen in osteopathia striata, and it is seen particularly well in the metaphyses and epiphyses of the long bones, the vertebrae and the tarsus. The hyperostosis, which on the whole is more pronounced on the external than the internal surfaces of the cortices, is seen on the shafts of all long bones. The density of the new bone varies considerably. In the metacarpals, metatarsals, and to a lesser extent the phalanges, there is no more than increased thickness of the dense cortices. In the major long bones the new bone is more variable in density though on the whole it is less dense than normal cortical bone and shows an irregular, fluffy surface. Some of these thickened bones are irregularly honeycombed, the appearance being rather suggestive of Paget’s disease. The hyperostosis of the long bones was more advanced and more universal in the patient reported by Uehlinger, possibly because he was twice the age of Dr Batten’s patient. The skull, ribs and vertebral bodies were comparatively free from cortical changes. In Uehlinger’s case there was fusion of the lateral articulations and ossification of the dorsal ligaments throughout most of the spine; the vertebral bodies showed well-marked vertical striation in both cases. The pituitary fossa appeared to be normal.

REFERENCES

CASE 5—OSTEOPATHIA STRIATA

(Figs. 13 to 17.) Male, first seen when twelve years of age. Always ran awkwardly: "something wrong" with right leg. At ten years developed rheumatic fever, which affected his heart; no definite arthritis at any time. Awkwardness of right leg more noticeable recently. Family history negative. On examination—no limp. Right thigh thinner than left; calves equal; right lower limb half an inch longer than left. Movement of all joints free. Heart: loud mitral murmur. Urine contained trace of albumin: no Bence-Jones albumose. Radiographs showed striation affecting particularly the right side of the pelvis, the right half of the sacrum, and the bones of the right lower limb. The bones of the right upper limb, including the glenoid and acromion, were also affected, but not to the extent seen in the lower limb. The bones of the right side were generally more translucent than those on the left, and this accentuated the striated appearance. There was a little mottling with dense spots in some of the epiphyses and in the cuboid. The shaft of the right fibula was thickened. The bones of the left upper and lower limbs were not entirely free from changes but these were much less marked than on the right side. There was one dense spot in the right capitate and some mottling of the right ischium. The skull showed nothing abnormal. Radiographs taken four years later showed even more marked striation, the bones on the right side still showing more advanced changes than on the left. Osteoporosis in the bones of the right side had practically disappeared. The right ischium showed an area of increased density: there were a few dense spots in the left ischium. At the age of twenty years there were no symptoms referable to the bone dysplasia.
FIG. 14
Case 5—Knee joints at the age of twelve years. Striation of the shafts and epiphyses is more marked on the right side. Right fibula is thickened.

FIG. 15
Case 5—Knee joints at the age of sixteen years. There is striation of both femora and tibiae. The shaft of the right tibia is now slightly enlarged as well as the fibula.
Case 5—Pelvis and femora at sixteen years, showing striation, more marked on the right side, and dense spots in the head of the right femur, the right ilium and in the left ischium. In the right ischium is a dense patch. Note that the osteoporosis on the right side is now much less obvious.

Fig. 16

Case 5—Ankles and feet at sixteen years. Striation is more obvious on the right, partly owing to the diminished density of the bones.

Fig. 17
CASE 6—OSTEOPATHIA STRIATA

(Figs. 18 and 19.) Boy, aged fourteen years. For three years he had complained of recurrent attacks of pain and swelling of the left knee joint; the attacks lasted from a few days to a week and then cleared up completely. At four years had scarlatina followed by “rheumatism”: recovery was complete. The blood and urine were normal. Radiographs showed marked striation of the metaphyses of all the long bones, the sacral wings, pelvis, patellae, calcanei, scapulae and the ribs near the costo-chondral junctions. There were also some clear areas and dense spots in the bones with slight enlargement of the metaphyses and a few small exostoses. His father, to a small extent, and his sister, showed similar changes (Dr Voorhoeve’s case).

FIG. 18
Case 6—Knee joint showing marked striation with slight enlargement of the metaphyses, and a local projection resembling an exostosis on the inner side of both femur and tibia (after Voorhoeve).

FIG. 19
Case 6—Pelvis and upper femora showing striation which radiates in the iliac bones. The necks of the femora are rather thick and both show a tiny spur projecting downwards (after Voorhoeve).
CASE 7—HYPEROSTOSIS GENERALISATA with striation of the bones

(Figs. 20 to 26.) Male, aged twenty-eight years. Admitted to hospital with pneumonia. Had always been healthy but underfed. Investigations into his history showed that there had been prolonged deficiency in calcium, and vitamins A, B₁, C and D. Probably a protein deficiency over a period of many years. Repeatedly unemployed. His father and an uncle said to be of same build as himself. Other members of family normal. Patient is well above the average in height and is thin. Blood examination—negative. Radiographs showed coarse striation of the ends of most of the long bones, and of the vertebral bodies, tarsus, sacrum and, less obviously, the pelvis and ribs. The hands and feet show increased thickness, with uniform density, of the cortices of the minor long bones. There is a varying degree of hyperostosis of the major long bones, and the normal contrast between cortex and medulla is obscured. On the whole the surface of the thickened bones is fairly smooth. The left tibia shows fusiform enlargement posteriorly in the upper half due to thickening of the cortex. Except for slight increase in density the skull shows no definite change. (By courtesy of Dr G. Batten.)
Case 7—The humerus shows altered texture and increase of density but no enlargement of shaft (Fig. 24). The forearm shows hyperostosis (Fig. 25). In the knee joint there is coarse striation and enlargement of metaphyses (Fig. 26).