A CASE OF OSTEOPATHIA STRIATA

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Osteopathia striata was the name applied by Fairbank (1935) to cases described by himself (1925) and by Voorhoeve (1924) and characterised by striation of the skeleton and particularly the metaphyses of the long bones.

CASE REPORT

A girl aged three years was first seen on account of nasal obstruction and recurring bronchitis. Birth had been difficult because of a large head, and delivery was completed with the aid of forceps. The neonatal period was stormy; she had pneumonia and feeding was difficult because of nasal obstruction. Some improvement took place after the first month, but the feeding difficulty persisted and she was referred, at the age of five months, to a London children's hospital. There it was noted that there was failure to gain weight and this was attributed to the considerable mechanical difficulty in feeding due to nasal obstruction. Thereafter gradual improvement took place, but she was late in passing all the milestones of infancy. Thyroid (half a grain twice a day) was given for a period of six months with further improvement in her general condition. She had pneumonia at the age of two years.

The parents were healthy and unrelated. There were two other children, girls aged five and a half and two years, who were normal in every way.

Examination—The weight and height were within the normal range for her age. The skull was large and brachycephalic; the circumference was $21\frac{3}{4}$ inches. There was no bossing and the fontanelles were closed. The eyes were widely set with a low forehead and the epicanthic folds were marked. The nose itself was narrow and there was marked obstruction with a purulent discharge. The mouth was constantly open. The tongue showed a marked median groove, the palate was highly arched and there was a bifid uvula. The teeth were all present except the left lower second molar. The hearing was normal to simple tests and the eye grounds were normal. There were several palpable cervical glands. The thyroid gland was normal.

The chest was somewhat barrel-shaped with a prominent sternum. The heart was normal. Both sides of the chest moved equally but the excursion was poor. There were signs of generalised bronchitis. The abdomen was distended and resonant to percussion. There was an umbilical hernia. The liver and spleen were not palpable. The vulva was normal. The stools were normal.

There was full mobility of all the joints and there was no deformity of the limbs or spine. The gait was normal. The muscles were normally developed and of normal tone but the power was weak in the limbs. The skin was normal.

The child was placid, but played and behaved normally. Speech was indistinct and had a marked nasal character. She could name familiar objects and all the letters of the alphabet.

Radiographic changes—Radiographs showed a fine striation of the long bones (Figs. 3, 4 and 5) which was more marked at the ends. The striation was linear, regular and vertical and was well shown in the lower femora and upper tibiae. The ilia (Fig. 2) showed a fan-like striation. The bones of the hands and feet were all within normal limits with the exception of the calcanea which showed a faint striation. The ribs, clavicles and vertebrae were normal. The skull was large and showed increased density of the base and the frontal bone (Fig. 1). Both antra and the right and left ethmoidal cells were obscured. The frontal sinuses had

FIG. 1

Skull showing increased density and thickness. This is well marked at the base.

FIG. 2

Ilia showing fan-like striation, especially in the left ilium.

FIG. 3

FIG. 4

Figure 3—Left humerus showing striation of the upper end. Right humerus was similar. Figure 4—Right radius showing striation of lower end. Left radius was similar.

FIG. 5

Lower femora and tibiae showing well marked striation.

not developed. No condensations or dense spots were seen. Radiographs of the other members of the family were all normal.

DISCUSSION

The widespread striation seen in this case is similar to that seen in the cases of Voorhoeve (1924), Fairbank (1925) and Hurt (1953). The striation is fine, linear and uniform and is most clearly seen in the growing ends of the long bones. It is well demonstrated in the lower femora and upper tibiae. The striation in the case reported by Fairbank (1925) is interesting in that it was predominantly unilateral. In the ilia the striation assumes a fan-like appearance; this was well marked in the case of Voorhoeve (1924). The cases of Fairbank (1925) and Hurt (1953) showed a more irregular fan-like appearance together with areas of increased density in the ilia.

The cases referred to above all showed areas of increased density in various bones such as the ilia, carpals and vertebrae. These areas of increased density, which are reminiscent of osteopoikilosis, are not seen in my case. Fairbank (1950) saw his patient some eight years after his original description and noted that more dense areas had appeared. It is possible that the age of my patient may account for their absence.

The radiographs of the skull were normal in the cases of Voorhoeve (1924) and Fairbank (1925). The changes in the skull radiographs in my case closely resemble those described by Hurt (1953). His case was that of a man of fifty-one years and the skull radiographs showed increased thickness and density of the cranial vault and base. All the sinuses, with the exception of the frontals, were obscured. No mention was made of any unusual facial appearance, nasal obstruction or defect of hearing.

No clinical picture has been associated with osteopathia striata because of the scanty signs. Voorhoeve's patients were a boy of fourteen years, his sister aged ten years and their father. The boy showed signs of mental retardation and muscular weakness. The case of unilateral striation, described by Fairbank (1925), was that of a boy of twelve years who showed some increase in length of the affected leg. Abnormal signs were more prominent in the present case: there were an unusual facial appearance, nasal obstruction, bronchitis and mental retardation.

Other cases (Busch 1937, Windholz 1932) have been recorded with striation and areas of increased density and some have been grouped with osteopathia striata (Lindbom 1942, Mascherpa 1931). But none showed the characteristic striation which was shown in Voorhoeve's (1924) cases.

Hurt (1953) discussed the relationship of osteopathia striata and osteopoikilosis. A relationship was suggested by Voorhoeve, but Fairbank (1950) maintained that they are two separate entities. The absence of dense areas in the bones of my patient, who is the youngest recorded with this affection, may support Fairbank's opinion. The relationship of osteopathia striata and osteopetrosis was also discussed by Hurt (1953) and he noted that the skull changes in his case were typical of osteopetrosis. The case of Fairbank (1948) which showed osteopetrosis of the upper femur and a fine striation of the lower end was quoted in support of this relationship. There is no doubt that striation is present in some cases of osteopetrosis, but it is usually in the form of streaks and is unlike the striation of osteopathia striata.

The differential diagnosis was fully discussed by Fairbank (1950) and Hurt (1953). There is, however, only one condition which can give rise to a widespread and uniform striation, and this is found in some cases of coeliac disease (Fairbank, personal communication).

It is clear that the term osteopathia striata can be applied to the present case and to those of Voorhoeve, Fairbank and Hurt. All these cases differed in many details but all showed the characteristic striation.

SUMMARY

A case of osteopathia striata in a girl aged three years is described. Similar cases are reviewed.

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