OSTEOPATHIA STRIATA—VOORHOEVE'S DISEASE*

REVIEW OF THE ROENTGEN MANIFESTATIONS

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OSTEOPATHIA striata is a rare roentgenographic entity characterized by fine linear striations seen primarily in the metaphyses and diaphyses of long bones in asymptomatic patients. The finding has been described in all bones except those of the skull and the clavicles. Patients in whom the entity has been described have no known associated physical abnormalities or characteristic laboratory findings.

Our purpose is to describe in detail the roentgenographic changes diagnostic of this disorder, to review the literature and to add 2 new cases. One of the cases is unique in that it is the only instance of unilateral skeletal involvement to be reported.

REPORT OF CASES

CASE 1. An 8 year old white girl has been followed for years at the Duke University Medical Center (DUMC) for speech therapy and mental retardation. The diagnosis of osteo-

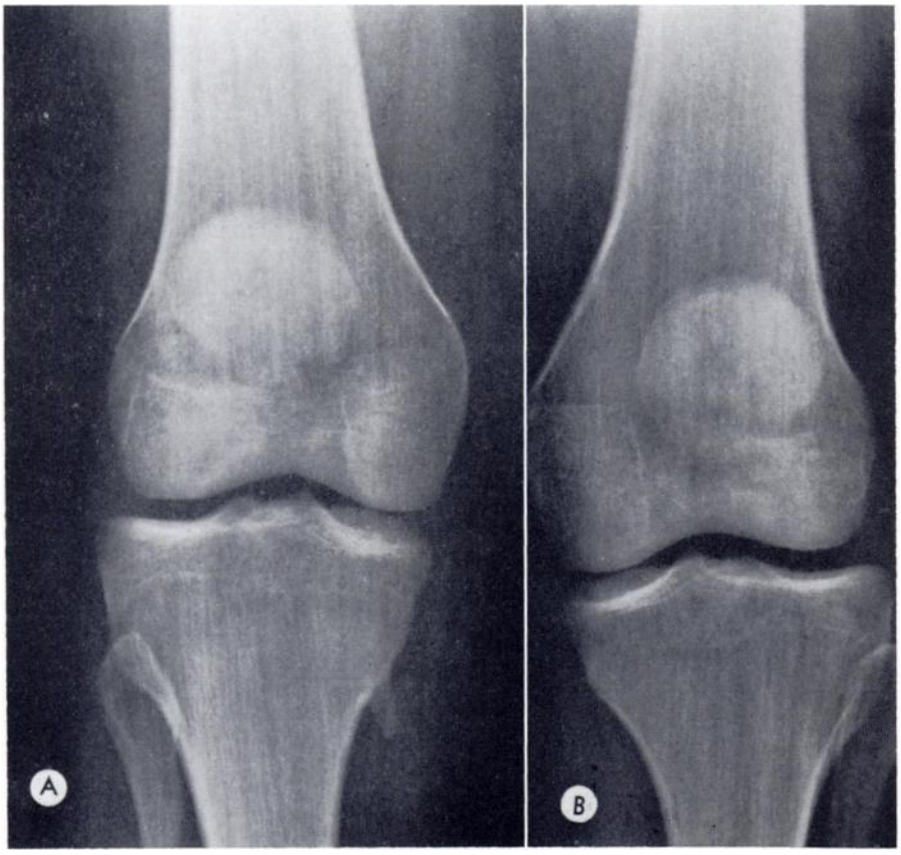

FIG. 1. Case 1. (*A* and *B*) Frontal roentgenograms of the knees at the age of 19 years. Linear striations in the distal femora and proximal tibiae parallel to the axis of the bone shafts. These changes are more prominent in the femora. Note tibial osteochondroma on the right (*A*).

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pathia striata was made at the age of 8 years when roentgenograms of the pelvis and knees were made because of vague left hip and leg pain. Incidental note was made of a small osteochondroma of the proximal right tibia.

Roentgenograms of the long bones taken at the age of 19 years show linear striations in the ends of the femora, tibiae and proximal humeri (Fig. 1, *A* and *B*; and 2). A small osteochondroma projects medially from the proximal right tibial metaphysis. Striations are also noted in the first metatarsals (Fig. 3, *A* and *B*). Views of the pelvis show striations in the iliac wings (Fig. 4), ischia and pubic rami. There are zones of bone condensation in the acetabular roofs and the humeral epiphyses. Changes of osteitis pubis are also present.

Case II. A 24 year old white woman was admitted to the psychiatric service of DUMC in May, 1971. All laboratory and physical findings were normal.

During her hospitalization she swallowed an open safety-pin. Plain roentgenogram of the abdomen, obtained to localize the pin, revealed abnormal striations in the right ilium. A subsequent bone survey showed unilateral changes of osteopathia striata limited to the right os coxa (Fig. 5), femur, tibia, fibula (Fig. 6, *A* and *B*), and calcaneus (Fig. 7, *A* and *B*). All other bones were normal.

HISTORIC REVIEW

The changes of osteopathia striata were first described by Voorhoeve, who reported 3 cases in 1924.[8] The entity went unnamed, however, until 1935, when Fairbank made the current designation.[3] Our review of the literature reveals a total of only 5 case reports,[1,2,8] but additional cases undoubtedly have been observed because the entity is illustrated in some of the standard textbooks[5,7] dealing with disorders of the skeletal system. However, the texts give no information about the patients.

ROENTGEN FEATURES

The diagnosis of osteopathia striata is based entirely on roentgen findings. No pathologic correlation exists because none of the patients described had a bone biopsy. The hallmark of the disorder is fine, dense, linear striations. In long bones, the striations are nearly uniform and are seen primarily in the metaphyses and diaphyses. They run parallel to the axis of the shaft of long bones and occasionally cross into the epiphyses. The striations apparently vary in length in direct relationship to the growth rate of the involved bone. The longest striations, therefore, are found in the femora. In the iliac wings, a fan-shaped pattern is seen (Fig. 4; and 5). This finding has been more striking in cases reported by others. Involved bones are normal in shape, density and cortical thickness.

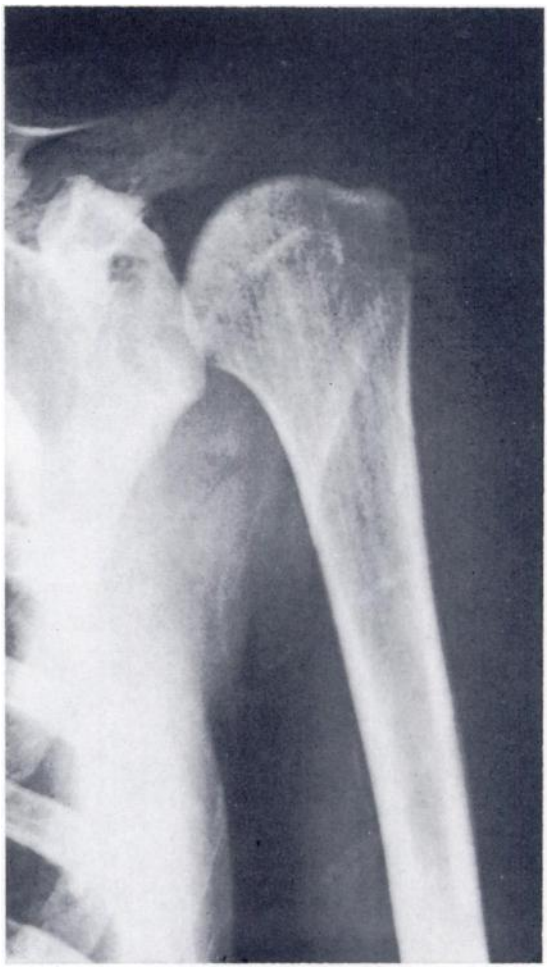

Fig. 2. Case I. Proximal left humerus in anteroposterior projection. Note that the striations are present in the epiphysis, metaphysis and diaphysis. A few areas of bone condensation are seen in the epiphysis.

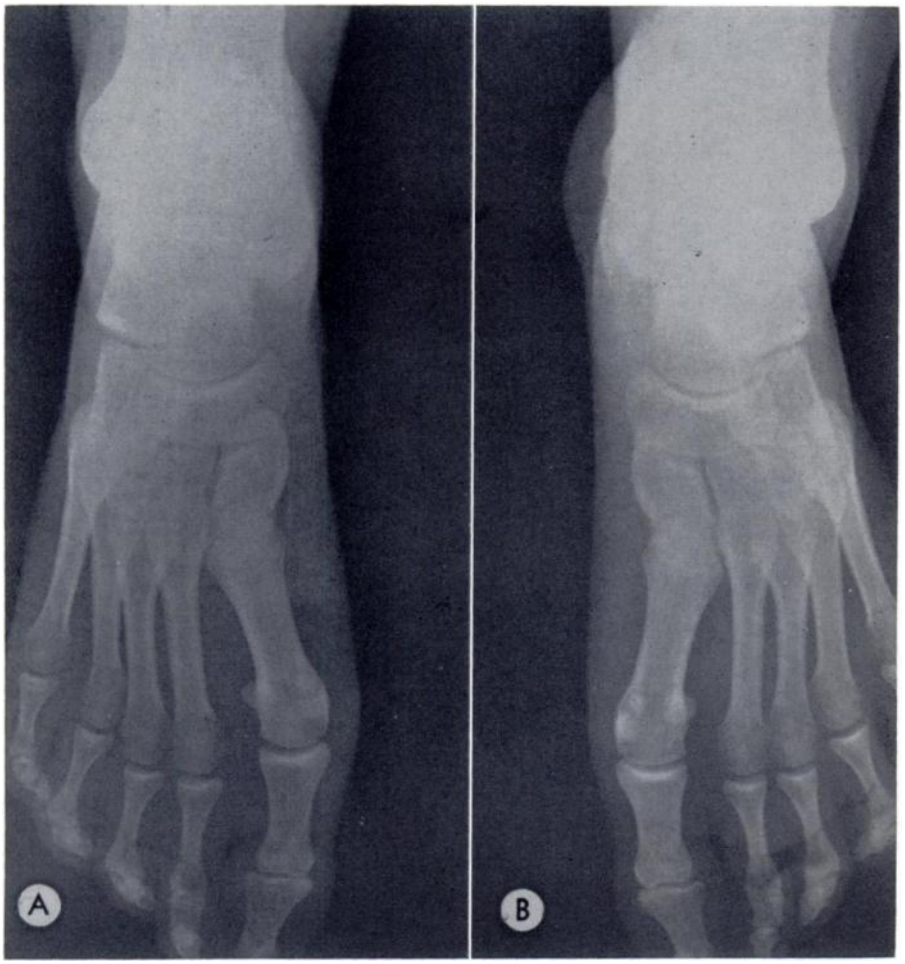

FIG. 3. Case 1. (*A* and *B*) Anteroposterior roentgenograms of the feet. Faint striations are seen in the first metatarsals, being more prominent on the left (*B*).

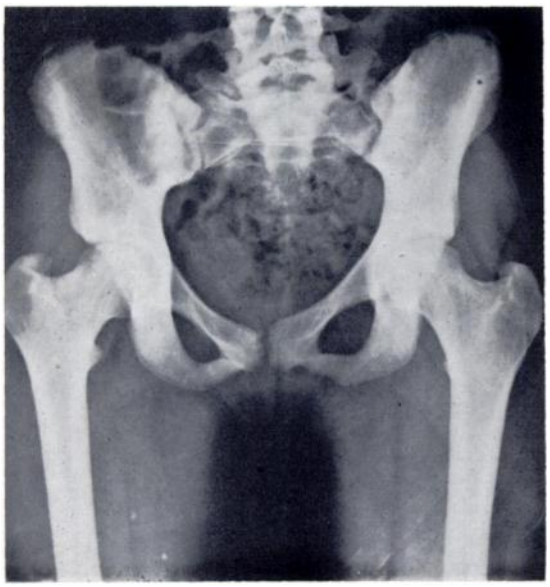

FIG. 4. Case 1. Anteroposterior roentgenogram of the pelvis and hips. Striations are fan-shaped in the iliac wings. Some striations are also seen in the pubis and ischium bilaterally. Both ischial tuberosities are more radiopaque than normal and there are zones of increased density in the acetabular roofs.

DIFFERENTIAL DIAGNOSIS

There have been reports of bone "striations" in other diseases, including osteopoikilosis, osteopetrosis, dyschondroplasia (Ollier's disease), melorheostosis, Paget's disease, polyostotic fibrous dysplasia and neurofibromatosis.[1,4,6] However, in all of these entities the striations are dissimilar to those found in osteopathia striata in

their location, length, width, or in the number of bones involved. Fairbank, in a personal communication to Bloor,[1] contended that the only other condition that "can give rise to widespread and uniform striation" is celiac disease. A brief review of the cases of celiac disease available to us failed to reveal any patients with bone striations. However, few of these patients had complete roentgenographic studies of their skeletal systems.

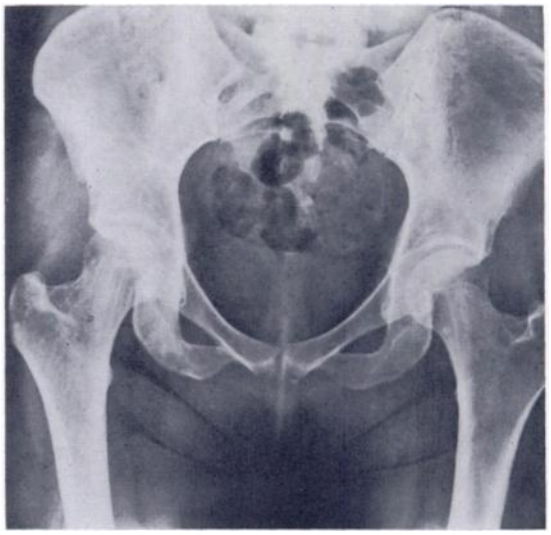

Fig. 5. Case II. Pelvis and hip joints at 24 years of age show striations on the right side in the iliac wing and proximal right femur. Zones of bone condensation are seen in the right acetabular roof.

DISCUSSION

The etiology and pathogenesis of this disorder are unknown. It is thought to be an error in modeling of bone and is classified by Rubin among the metaphyseal dysplasias.[7] However, the author states clearly that it does not fit his classification of bone dysplasias.

Four of the 7 patients whose cases have been reported were females. The only possible hereditary or familial influence was present in the 3 cases reported by Voorhoeve,[5] which included a father, son, and daughter who showed similar bone changes.

In all patients except our Case II, in-

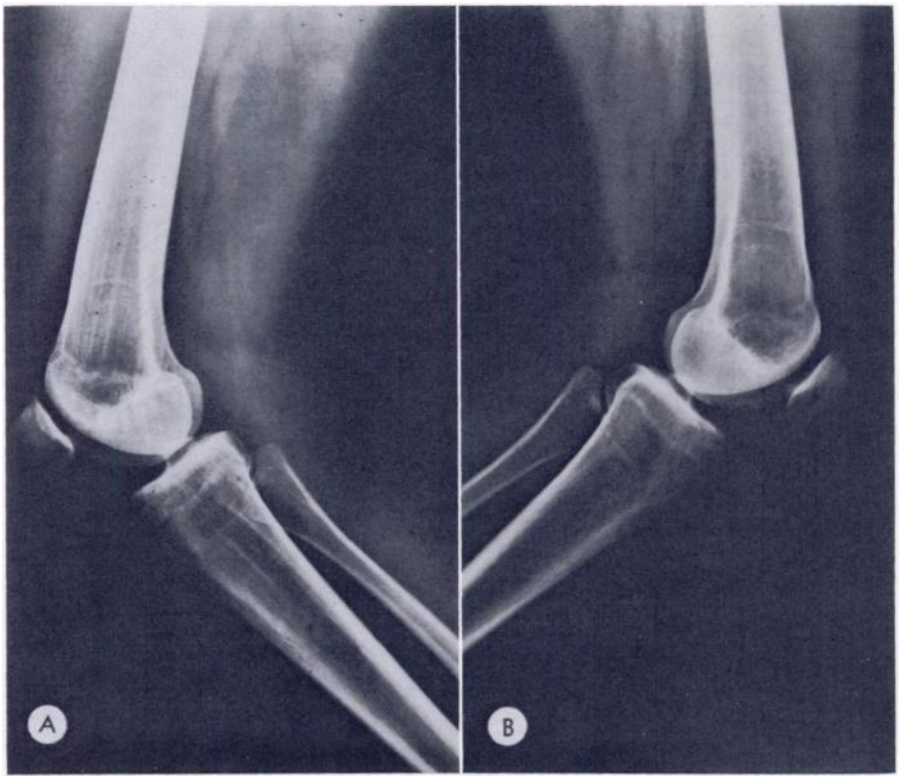

Fig. 6. Case II. (*A* and *B*) Lateral roentgenograms of the knee joints show striations in the femur, tibia and fibula on the right. The left side is normal (*B*).

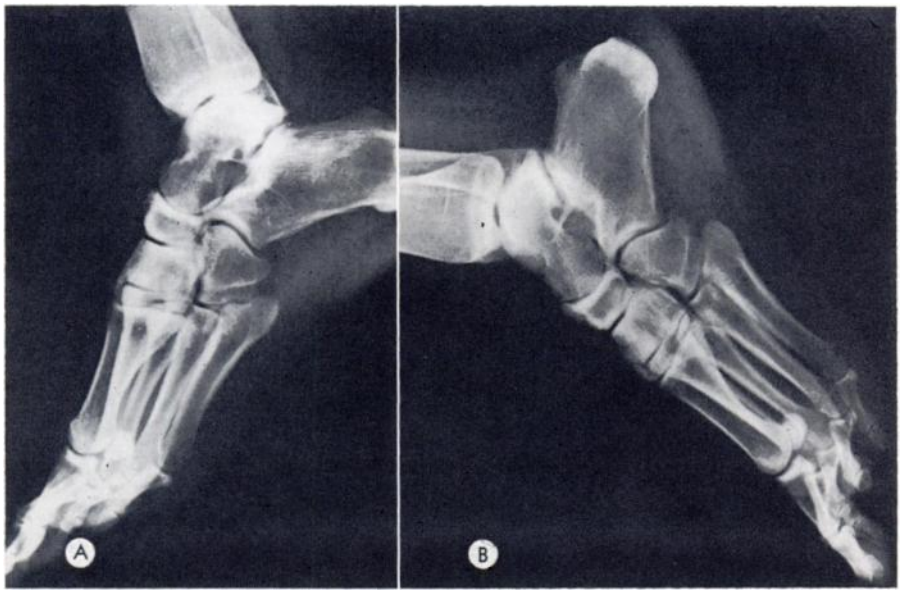

Fig. 7. Case 11. (*A* and *B*) Lateral roentgenograms of the feet. Faint linear striations are present in the calcaneus on the right as well as in the distal right tibia (*A*).

volvement was bilateral. Fairbank, in his original paper,[3] also reported a case of unilateral involvement in a 12 year old boy, but striations eventually became apparent bilaterally and the author corrected his findings in a later report.[4]

Several authors have postulated a relationship with other diseases. Voorhoeve believed that the entity was a congenital defect related to dyschondroplasia and osteopoikilosis. Fairbank stated that the entity could not be considered a form of dyschondroplasia because the changes in the hands and feet in that disorder could not be considered striations and there was no uniform involvement of the major metaphyses—as in osteopathia striata.

Hurt[6] suggested that osteopathia striata may be a form of osteopoikilosis in which the striations predominate. Evidence for this theory is the finding in 6 of the 7 reported cases of bone condensations somewhat suggestive of osteopoikilosis. Hurt speculated that force applied to long bones involved by the underlying entity could produce changes typical of either disorder. Fairbank[3] agreed that occasional streaking could be seen in osteopoikilosis, but he pointed out that these streaks were irregular and shorter than those found in osteopathia striata.

Hurt[6] also raised the question of a relationship to osteopetrosis. One of his patients showed changes typical of that entity in the skull and ribs and changes of osteopathia striata in the long bones. His case resembled that reported by Bloor[1] in which the patient had bone sclerosis at the base of the skull and thickening of the frontal bone. This patient did not have the bone condensations seen in the other 6 cases reviewed here.

Thus, it can be seen that there has been a significant debate and postulation about the nature of this entity and the underlying disorder, but no definitive information has become available. Since patients showing the skeletal changes of osteopathia striata have no clinical or laboratory abnormalities, there is no indication for bone biopsy or other more aggressive diagnostic maneuvers that might help to explain the pathogenesis. Failing this, the disorder remains a purely roentgenographic entity.

SUMMARY

Osteopathia striata is a rare skeletal disorder manifested roentgenographically by fine, linear striations primarily in the long bones.

There are no associated clinical or laboratory abnormalities.

A review of the literature reveals only 5 reported cases.

Two new cases, including 1 with unique unilateral involvement, are reported.

Several hypotheses about the etiology of the disorder are discussed.

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