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Idiopathic Juvenile Osteoporosis:

Report of a Case and Review of the Literature

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A case of idiopathic osteoporosis involving the whole vertebral column of a girl 10 years of age is described. The condition subsided spontaneously after four years. The clinical, biochemical and radiographic aspects as also the therapeutical possibilities of idiopathic osteoporosis are discussed on the evidence of the 18 cases reported in the literature.

Osteoporosis in childhood is usually a sequel to some primary disease. Most forms of the condition are well-known from the pioneering investigations of ALBRIGHT and REIFENSTEIN [1, 13], and others. Juvenile osteoporosis may follow immobilization when normal osteoblastic activity lacks stimulation; it may further be associated with Cushing's disease irrespective of this being spontaneous or induced by hormone therapy: excessive amounts of glucocorticoids inhibit the formation of bone matrix, a part phenomenon of general protein antianabolic activity. Osteoporosis may moreover accompany gonadal dysgenesis, thyrotoxicosis, starvation, steatorrhoea, vitamin D deficiency (osteomalacia), hyperparathyroid fibrous osteitis, and it is justified to classify osteogenesis imperfecta also under the heading of congenital osteoporosis [8].

Several reports have been published in recent years on cases of osteoporosis of unknown aetiology [2, 5, 14, 6, 12]. Their data are assembled in Table I.

REPORT OF A CASE

E. K., a girl of 10 years, had fractured her leg at the age of 2, and had had nephritis at the age of 4 years. During the last year she had put on much weight. Three months before admission she had a fall and has had back pain since then. She was referred to us in 1964 with the suspicion of Cushing's disease. At admission, physical and laboratory examination including renal clearances, urinary output of 17-ketosteroids and 11-oxysteroids yielded normal results. Serum calcium was 10.6 mg per 100 ml; serum phosphorus, 4.6 mg per 100 ml; alkaline phosphatase, 8 B. U.; Sulkovitch's test for Ca in urine was negative. X-rays revealed marked osteoporosis involving the thoracic and lumbar vertebral bodies ("Fischwirbel"). The spongiosa appeared to be vitreous, transparent, with a blurred structure. The cortical pattern was sharp, the intervertebral disks were swollen. But for a moderate porosity, the other bones showed no changes (Fig. 1).

Attempts to ascertain the disease responsible for the osteoporosis were unsuccessful. The patient was kept under observation for half a year; on anabolic hormone and oral calcium treatment there was a slight radiographic improvement and a gradual subsidence of the dorsal pains.

TABLE I

Cases of idiopathic osteoporosis

No.	*Reference*	*Sex,*	*Age yrs*	*Radiographic symptoms*
1.	(2)	♂	12	O. of leg and pelvis
2.	(2)	♂	12	Compression of thoracic and lumbar vertebrae
3.	(3)	♂	13	O. of vertebral column. Biconcave verterae
4.	(4)	♂	11	Extensive O. Vertebral compression, multiple fractures of long bones
5.	(4)	♀	8	Extensive O. Vertebral compression, fracture of long bone
6.	(4)	♀	9	Extensive O. Vertebral compression, fracture of long bones
7.	(4)	♂	9	Extensive O. Vertebral collapse
8.	(4)	♂	11	Extensive O. Vertebral collapse, healed costal fracture
9.	(4)	♀	9	Extensive O. Flattened vertebrae
10.	(5)	♂	12	O. of vertebral column, biconcave vertebrae
11.	(8)	♀	11	O. of vertebral column, biconcave vertebrae
12.	(9)	♂	16	General O. Fracture of right femur
13.	(9)	♀	16	Extensive O. Fracture of right tibia
14.	(10)	♀	7	O. of vertebral column, biconcave vertebrae
15.	(11)	♂	15	Extensive O. Biconcave vertebrae, fracture of long bones
16.	(11)	♀	12	Extensive O. Vertebral compression, fracture of sternum and knee
17.	(12)	♀	12	Extensive O. of vertebral column, vertebral compression
18.	(14)	♀	11	O. of vertebral column, biconcave vertebrae, fraction of femur
19.	present case	♀	10	O. of vertebral column, biconcave vertebrae

At a follow-up examination four years later, at the age of 14 years, X-rays revealed vertebral bodies of normal shape and height, further a slightly rarified and somewhat thickened trabeculation. The diminution of physiological curvatures had led to the development of a flat dorsum (Fig. 2). The girl had grown 27 cm and gained 23.5 kg during four years. She is now doing physical work as an industrial apprentice. Repeated laboratory tests were invariably normal. The case was classified as one of idiopathic juvenile osteoporosis with practically spontaneous recovery.

reported in the literature

Clinical symptoms	*Ca balance*	*Treatment*	*Results*
Pain in leg	negative	Ca, vitamins, anabolic steroid	No change
Back pain	—	Anabolic steroids	Symptomatic improvement
Back pain	—	—	Recovery
Repeated fractures	negative	Dihydrotachysterol, sex hormones	Crippled
Generalized pain	positive	Dihydrotachysterol, sex hormones	Improvement. Normal X-rays of vertebral column. Low stature
Progressive pains	negative	Triparanol, dihydrotachysterol, sex hormones	Gradual improvement
Back pain	positive	—	Unexpected clinical and radiological improvement
Pain in hands, legs and back	negative	Dihydrotachysterol, Ca	Marked clinical and radiological improvement, normal height
Pain in back, right knee and leg	doubtful	—	Clinical improvement
Back pain	—	—	Recovery
Back pain	—	—	Recovery
Pain in back and extremities	negative	Ca, anabolic steroids	Practically recovered
Pain in back and extremities	negative	Ca	Practically recovered
Back pain	—	—	Recovery
Pain in back and leg	—	Physicotherapy, anabolic steroids	Marked clinical and radiological improvement
Back pain	—	—	Recovery
Back pain	negative	—	Recovery
Pain in back and extremities	negative	Ca; vitamin D, Anabolic steroids, Ca.	Moderate improvement Recovery
Pack pain	—	—	Spontaneous recovery

The 18 cases described in the literature [2, 3, 4, 5, 8, 9, 10, 11, 12, 14] present the uniform picture of a well circumscribed disease entity starting during or immediately before puberty.

Clinical aspects. The disease affects both sexes and appears between 9 and 11 1/2 years in girls, between 9 and 15 years in boys, with back pain as the predominant symptom. The pain which at first is uncertain is seldom so

FIG. 1

grave as to cause a radiographically visible affection of the vertebral column or a compression fracture of the vertebrae. The ESR may be elevated but soon returns to normal. Clinical examination excludes the possibility of any other disease. Osteoporosis of the extremities may be complicated by their fracture. It is the vertebral column which shows the gravest alteration, while porosity of the other bones may be slight or pronounced.

X-ray aspects. Changes in the vertebral bodies may involve their inner structure or their whole aspect. The spongiosa is homogeneous, transparent, the trabecules are indistinguishable. The vertebral surfaces are so sharply outlined as if they had been "traced by Indian ink". Diagnosis on the strength of this picture is difficult because differences in X-ray technique and in the thickness of the soft tissues may cause extreme variations.

Swelling of the intervertebral discs results in a biconcavity and possibly also a discontinuity of the surfaces, a tapering and a more or less marked compression of the vertebral bodies. As in all forms of osteoporosis, the other bones may also be involved. A diffuse change of the vertebral column is the decisive morphological feature.

Biochemical changes. As in the other forms of osteoporosis, the condition is not associated with haematological changes. Hypercalciuria exists only during the development and progression of the disease. Since patients solicit medical aid only at the appearance of the first symptoms, at a time when osteoporosis had reached a peak and a new equilibrium has been established, urinary calcium output is no longer elevated: in other words, at the time of hypercalciuria osteoporosis is radiologically not yet perceptible. At this time, faecal excretion of calcium exceeds its oral intake; the organism is unable to compensate the deficit by a corresponding reduction of renal calcium excretion so that a negative Ca balance will result, although excess amounts of Ca are needed for the growth of children.

Biopsy. Since osteoporosis consists in the atrophy of the proteinaceous matrix, it shows no striking symptoms. The trabecules are thinner and less than normal, and the number of osteoblasts is diminished. These quantitative changes can be appreciated only by comparison with normal individuals of identical age.

Course of the disease. The condition shows a tendency to spontaneous recovery. The responsible factors are obscure. After some years the Ca balance becomes positive and the vertebral bodies display signs of regeneration. The latter occurs in two ways. (i) The thinned trabecules undergo remineralisation. Completely decomposed trabecules are not remineralized. (ii) Healing is promoted by the considerable growth capacity of the still immature skeleton. As in the case

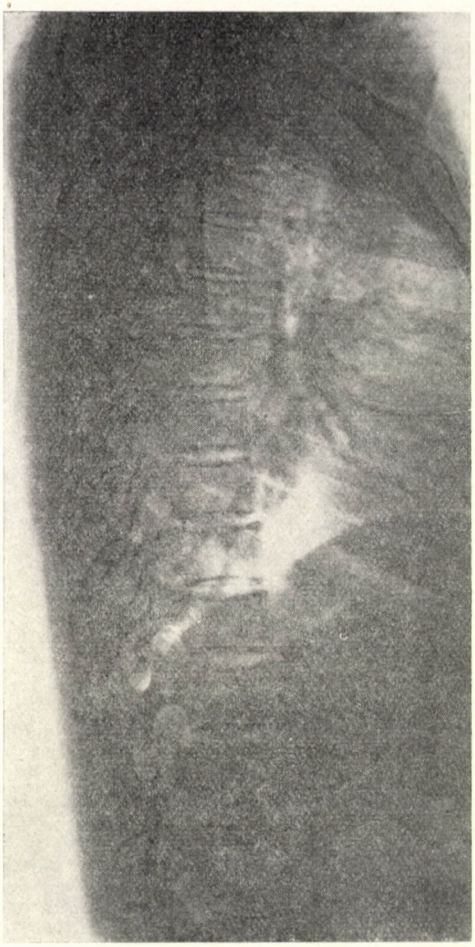

Fig. 2

of normal growth, the vertebral bodies are reconstructed by appositional growth on their surfaces cranio-caudally and ventrally. Growth in such cases is more intensive than in healthy children of the same age. Although the trunk, too, shows a higher than normal rate of growth during the period of recovery, the height of such children still remains under the mean.

Improvement, as shown by the X-rays, is actually the spontaneous correction of a defect which leaves the child with a disposition for orthopaedic disorders [6].

The *aetiology* is obscure. DENT and FRIEDMAN [5] registered a negative calcium balance with strongly increased faecal Ca excretion; the intestinal loss of Ca was, however, presumably not the cause but the consequence of osteoporosis. The disease failed to respond to androgens and oestrogens, but clinical improvement ensued in the majority of their cases including two untreated ones.

The sudden onset of the disease and the reversibility of demineralization are suggestive of osteoporosis of the Cushing type. One might think in this case of dealing with a monosymptomatic form of Cushing's disease lacking the symptoms of adrenal hyperfunction. Investigating a case of this kind, FANCONI et al. [6] found a normal 17-ketosteroid excretion. DENT and FRIEDMAN [5] observed a case in which the amount of excreted 17-ketosteroids increased on the administration of ACTH; adrenal biopsy revealed a normal histological pattern. Paper chromatography of the urine showed abnormal steroid metabolites in three patients. Although supporting data in this respect are scanty, the possibility of a transitory disturbance of steroid metabolism during puberty cannot be excluded.

As to *differential diagnosis*, the possibility of primary hyperparathyroidism should always be considered in cases of acute osteoporosis. The blood shows, however, characteristic changes in such cases and the X-ray symptoms (cysts, erosions, expansion of bones), too, are different from those seen in simple porosity. In a case of this kind, four parathyroid glands of normal size and histology were found [5].

Diagnosis is difficult in mild cases of imperfect osteogenesis as the symptoms of the latter condition sometimes become manifest only in puberty or in early adulthood. CLOUTIER et al. [4], employing YOWSEY's method, carried out quantitative microradiographic examinations in a case and found no changes that would have pointed to either imperfect osteogenesis or osteomalacia. Comparison with normal individuals of equal age showed increased bone reabsorption and decreased bone formation. After clinical recovery and normalization of the X-ray picture, biopsy revealed normal bone reabsorption and formation. Malnutrition, malabsorption, prolonged immobilization or other diseases predisposing to osteoporosis were absent in all of their patients.

Thus, the diagnosis of idiopathic osteoporosis depends on the following criteria:

(i) pronounced demineralization in the vertebral column;

(ii) absence of skeletal curvature, local erosions or cysts; and an unimpaired lamina dura of the alveolar processes;

(iii) no changes in blood chemistry;

(iv) normal urinary tests except for occasional hypercalciuria.

(v) no prolonged immobilization, atrophy, or steatorrhoea;

(vi) even if all these requirements are satisfied, diagnosis should be set up by the method of elimination, and the first diagnosis cannot be but conjectural.

Treatment. The tendency of the condition to spontaneous improvement makes it difficult to estimate the therapeutic value of anabolic steroids, vitamin D, oral calcium treatment, and of other methods.

The patient in the evolutionary phase of osteoporosis should sleep in a plaster cast and during the day wear a supporting splint. Adequate physicotherapy has the purpose to prevent accessory osteoporosis caused by immobilization.

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