Hyperparathyroidism Associated with Rickets

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SUMMARY

A 15-year-old Black patient was found to have primary hyperparathyroidism associated with rickets — an exceedingly rare feature. The possibility of nutritional rickets associated with tertiary hyperparathyroidism is discussed. Healing of rickets occurred after parathyroidectomy and administration of vitamin D with calcium supplements.

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Rickets associated with primary hyperparathyroidism is exceedingly rare. We could find only 2 cases of rickets caused by or associated with primary hyperparathyroidism.[1,2] Nutritional rickets associated with tertiary hyperparathyroidism has, to our knowledge, not been previously described.

This report describes a case of rickets associated with autonomous hyperparathyroidism which could be primary or tertiary.

CASE REPORT

A Black male, aged 15 years, was admitted with a history of being unable to walk for many years. His diet did not include adequate amounts of milk products and meat, although he was exposed to solar irradiation. Clinically he was a dwarf only 130 cm in height. He had the bony deformities of rickets, viz genu valgum, rickety rosary and Harrison's sulcus.

Investigations showed that the serum calcium was 11,0 mg/ml, serum phosphorus 2,4 mg/100 ml, serum alkaline phosphatase 124 King-Armstrong (KA) units, serum sodium 140 mEq/litre, serum chloride 110 mEq/litre, and serum potassium 5,0 mEq/litre. A Schilling test, vitamin A absorption test, barium meal and glucose tolerance test to exclude malabsorption, were all normal. The blood urea was 13 mg/100 ml, and the glomerular filtration rate (radioactive chromium EDTA) was 112 ml/minute. The urine showed gross panamino-aciduria, with no glycosuria. The NH_4Cl test, done according to the short method of Wrong and Davies,[3] showed that the kidneys could not acidify urine, in that the pH of the urine was 6,2 in the presence of systemic acidosis. The phosphorus excretion index was 0,432 (normal +0,09 to −0,09). Intravenous pyelography was normal.

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Radiographs of the skeletal system showed a pelvis with a triradiate appearance and radiolucency of the bones. There was evidence of rickets in both wrist and knee joints, with genu valgum of the knees (Fig. 1). The skull showed generalised loss of bone density with loss of lamina dura of the teeth; and pseudo-fractures (Looser's zones) were present in the lateral ends of the clavicles. The spine showed loss of bone density with marked widening of the intervertebral discs; in addition, increased density of the superior and inferior margins of the vertebrae was present ('rugger-jersey' spine) (Fig. 2). The hands showed evidence of hyperparathyroidism in that there was subperiosteal bone resorption of the phalanges.

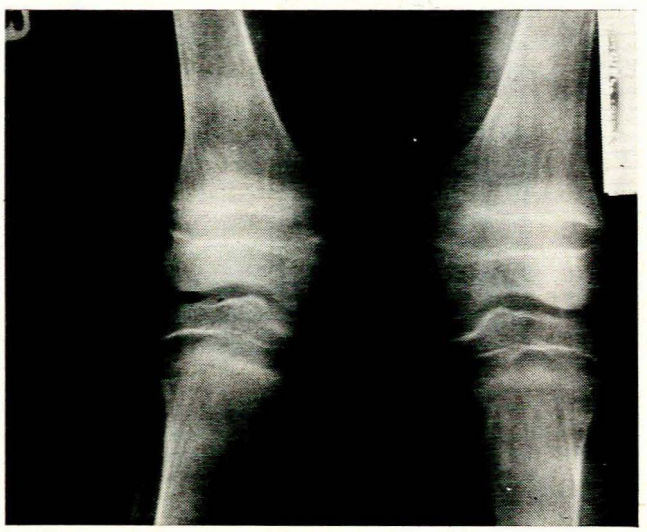

Fig. 1. X-ray film of knee joint showing the changes of rickets.

The patient absconded and was readmitted on 20 August 1973 in a bed-ridden state. The skeletal deformities were unchanged. Further investigations during this admission showed that the serum calcium was 11,4 mg/100 ml, serum phosphorus 2,7 mg/100 ml and serum alkaline phosphatase 124 KA units. The radiological changes were the same as on the first admission. A therapeutic test of vitamin D 2 000 units orally was given and after 2 weeks the serum calcium rose to 14,4 mg/100 ml. It was therefore decided to explore the patient's parathyroid glands surgically.

The neck was explored through a low transverse incision. A large parathyroid adenoma, 3 × 1,5 cm, was found in the position of the left upper parathyroid gland. Three other normal-looking parathyroid glands with an estimated total mass of 30 mg were identified and histological confirmation of their nature was obtained. The

parathyroid adenoma was excised *in toto*. Histological section of the parathyroid adenoma demonstrated masses of chief cells; oxyphil and water-clear cells were also present. There was no evidence of malignancy.

After surgery the patient had manifestations of hypocalcaemia with positive Chvostek's and Trousseau's signs. The serum calcium was 5,6 mg/100 ml and the electrocardiogram showed a corrected Q-T interval of 0,42/sec. He was treated with vitamin D 50 000 units daily and calcium supplements. The serum calcium rose to 8,4 mg/100 ml, serum phosphorus 4,4 mg/100 ml and there was no amino-aciduria. The urine osmolality rose to 863 milli-osmole/kg while the serum osmolality was 276 milli-osmole/kg. The kidneys' capacity to acidify urine returned. A renal biopsy showed normal renal histology. The patient became ambulatory and 3 months later a skeletal survey showed healing of the rickets with remineralisation of the bones (Fig. 3).

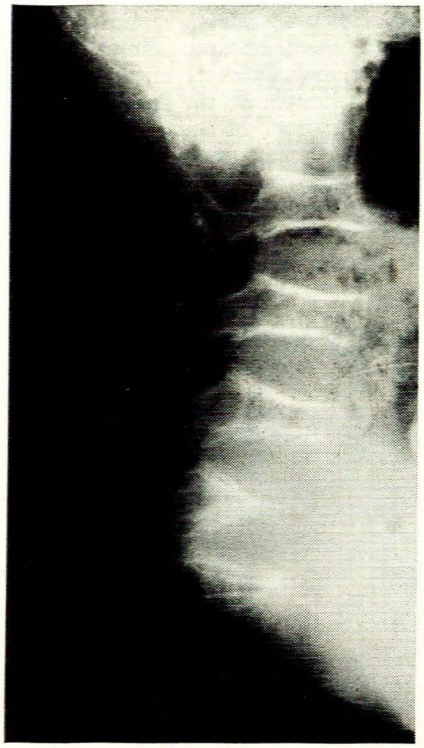

Fig. 2. X-ray film of spine showing sclerosis of superior and inferior margins of the vertebral bodies with radiolucency of the bone and marked widening of the intervertebral disc.

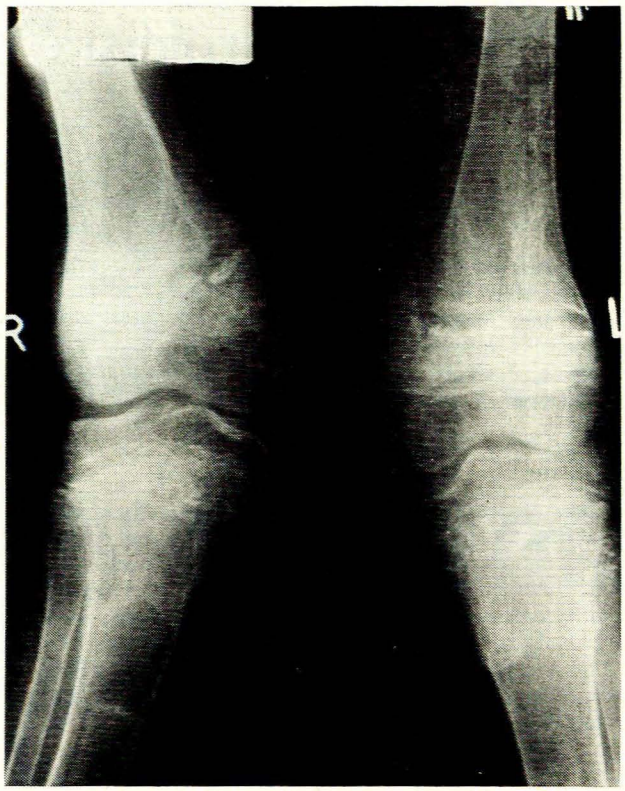

Fig. 3. Metaphyseal calcification after parathyroidectomy and vitamin D administration.

COMMENT

Our patient was originally diagnosed as a case of Fanconi syndrome because of the gross amino-aciduria and phosphaturia. However, this diagnosis was incorrect because the panamino-aciduria disappeared after parathyroidectomy and administration of vitamin D in large doses. Moreover, renal biopsy showed normal histology. The gross panamino-aciduria was due to deficiency of vitamin D, which is necessary for the reabsorption of amino acids.[4]

There are only 2 reported cases of rickets associated with hyperparathyroidism.[1,2] In the patient of Sano *et al.*,[2] although rickets and biochemical evidence of hyperparathyroidism were present in an 11-year-old boy, the patient was not subjected to surgery and thus definite evidence of hyperparathyroidism was not obtained. Woodhouse *et al.*[5] treated 2 cases of primary hyperparathyroidism and vitamin D deficiency with small doses of vitamin D and found that remineralisation of bones occurred. They suggested that vitamin D deficiency is responsible for the development of bone disease in some patients with hyperparathyroidism, and that there may be an increased requirement for vitamin D in this disorder. We therefore decided to treat our patient with small doses of vitamin D initially, but because the serum calcium rose to 14,4 mg/100 ml the patient was subjected to parathyroidectomy. It is possible that vitamin D deficiency was responsible for the rickets in our patient, as a result of increased requirements of vitamin D because of primary hyperparathyroidism.

Primary hyperparathyroidism is extremely rare in children, and in 1960 Nolan *et al.*[6] collected 22 cases from the

literature and described a further case. Because primary hyperparathyroidism associated with rickets is very rare it is possible that our patient had nutritional rickets with tertiary hyperparathyroidism. Nutritional rickets or osteomalacia is rare, and dietary sources of vitamin D are only required when man is shielded from effective sunshine by clothing, housing conditions or industrial smog.[7] It is possible that our patient had nutritional rickets due to dietary lack of vitamin D with tertiary hyperparathyroidism, even though exposed to solar irradiation. Seedat and North-Coombes[8] have observed adult nutritional osteomalacia in Blacks in Southern Africa. If this theory is tenable then this is the first case, to our knowledge, of nutritional rickets producing tertiary hyperparathyroidism.

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