Spinal Cord Compression Caused by Vitamin D Resistant Rickets

Case Report

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Vitamin D resistant rickets has for the most part been a concern of the internist, pediatrician or orthopedist. The neurosurgeon usually sees these patients only in the evaluation of craniosynostosis which is said to occur more frequently in patients with Vitamin D resistant rickets than in the general population. \cite{4,7} We have been unable to find reports indicating that the bony changes of Vitamin D resistant rickets may be a cause of spinal cord compression.

We have studied an adult who had the stigmata and family history typical of Vitamin D resistant rickets and who also had progressive paraplegia which we believe was caused by abnormalities of the lumbar and thoracic vertebrae secondary to this disease.

Case Report

A 60-year-old man had a family history compatible with a sex-linked dominant pattern of Vitamin D resistant rickets. One year before admission he had noticed numbness slowly advancing from the right foot to the level of the mid-abdomen. Over the ensuing months similar loss of sensation developed in the left leg and foot. The sensory loss was more severe distally than proximally. There had also been slight weakness of both legs for a year, with flexor spasm for the last 3 months. Six weeks before admission, the patient fell and for a few hours could not move his foot or legs. Afterwards the flexor spasm of the legs increased greatly in severity, and he could stand and walk a few steps only with help. Four weeks before admission the patient stopped trying to walk, mainly because of the spasms. He had noticed intermittent mid-lumbar pain after his fall but with no extension into the legs. This pain was not aggravated by coughing or sneezing. He had no sphincteric disturbance, except for slight hesitancy with urination, noticed first some time after the fall.

The family history was compatible with sex-linked dominant Vitamin D resistant rickets. Eight members of the patient’s family over the last 4 generations had been small of stature and had bowed legs.

Examination. The patient was 5 ft. 2 in. tall and had severe bowing of the legs. The bony configuration of his head was normal. The general physical examination otherwise was normal. There was a sensory level at T-10 with some preservation of gross touch and pin prick below this level. Sense of passive movement was lost in the toes and feet. All muscle groups in the legs were weak with the proximal groups being affected more than the distal groups. Muscle tone was greatly increased in the legs. The knee jerks and ankle jerks were hypertensive and both plantar responses were extensor.

Laboratory Data included the following: serum calcium 10.2; phosphorus 2.3; alkaline phosphatase 8.4; acid phosphatase 3.3. The cerebrospinal fluid showed 403 mgm. of protein, and a colloidal curve of 1111210000.

X-rays of the skull and chest were normal. X-rays of the thoracic spine showed narrowing of the disc spaces at most levels and spur formation in the mid-thoracic vertebrae. The interpedicular distance at T-11 and T-12 was reduced. The lumbar spine showed bi-concave vertebral bodies and bony proliferation in the anterior longitudinal ligament (Fig. 1). In x-rays of the left hip there was an irregularity of the density and contour of the femoral neck, and the proximal part of the femur was bowed with thickening of the cortex medially (Fig. 2).

A myelogram demonstrated subtotal obstruction at T-13 with the main compression being posterior. This was interpreted as consistent with an extradural obstruction (Fig. 3).

Operation. The L1 spinous process and lamina seemed to be fairly normal, but the spinous processes at T-12 and T-11 were extremely thick and hard. Also, at these levels, there was a great overgrowth of bone around the articular facets. The spinal canal beneath both of these vertebrae was greatly reduced in size by the overgrowth of bone. There was no epidural space, and the dura was closely approximated to the bone, and in certain areas adherent to it. The laminectomy was difficult because of the thickness and hardness of the bone and the reduced size of the spinal canal which was snuggly filled by the dura and cord. The adherent dura was torn in several places. Because of the hazards of persisting with the laminectomy and the total uncertainty as to how far the osseous changes might continue in a rostral direction, the procedure was terminated.

Postoperative Course. There was a rapid and uneventful recovery from the operative procedure. There was no objective neurological improvement, but the patient said that he had some decrease in the numbness. He has been seen several times since his discharge and has shown no change in the neurological status.

Discussion

Vitamin D resistant rickets or “phosphorus diabetes” was first described by Albright et al. in 1936.\cite{1} The osseous changes are similar to those of Vitamin D deficient rickets but do not respond to Vitamin D unless massive doses are given.\cite{2,4,8}

Many studies concerning the genetic transmission of this type of rickets have been published. The disease is transmitted as a sex-linked dominant trait but only sporadic cases have been re-
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Fig. 1. X-ray of thoracic spine showing spur formation and narrowing of the disc spaces at most levels.

Fig. 2. X-ray of left hip showing irregularity of the density and contour of the femoral neck.

ported. Metabolic studies in children with Vitamin D resistant rickets have shown a hypophosphatemia with normal serum calcium and elevated serum alkaline phosphatase. Adults usually have only a hypophosphatemia.

There is still no unanimity concerning the etiology of the disease. Albright et al. said that it represents an expression of secondary hypoparathyroidism caused by a decreased absorption of calcium from the bowel. Pierce et al. thought that the disease is caused by a tubular wasting of phosphorus resulting from a genetically or otherwise acquired defect in tubular enzymatic mechanism which retrieve phosphorus from the glomerular filtrate.

Long-term studies of children with Vitamin D resistant rickets show that when they become adults, most of them have retardation of growth and deformities mainly of the long bones that support the body weight. The exact factors limiting normal body growth are not completely understood, but deformities probably constitute the most important factor. In addition, a genetic nutritional, or direct growth-retarding factor may be present. Hypophosphatemia by itself has been shown to retard normal growth. The deformities which have been described most frequently are increased antero-posterior diameter of the skull with frontal bossing and bowing of the tibia and femur. The surgical correction of these deformities has been the topic of numerous publications.

X-rays of the bones of patients with Vitamin D resistant rickets show the same changes in the epiphysis and metaphysis which are seen in patients with Vitamin D resistant rickets who are in an active phase of the disease. The bony changes resulting in the previously described deformities are also seen. X-rays of the lumbar spine have revealed the type of changes and bi-concave vertebrae, which were seen in our patient.

The complication of spinal cord compression secondary to the changes in the bony vertebrae...