

## Surgically Curable Hypophosphatemic Rickets

All patients with apparent hypophosphatemic rickets (HR) do not have an inherited defect. A small but significant proportion have a tumor, which results in a very similar clinical picture. The history of one such patient and a review of the literature comprise the contents of this article.

An 8-year-old boy with rickets had swollen wrists for 6 months and knee pain for 30 months. His height had continued between the 25th and 50th percentiles. The only physical findings were tenderness and swelling of the wrists and right knee and genu valgum.

The findings were consistent with the proposed diagnosis of HR. Roentgenography confirmed the diagnosis. Demineralization of the pelvis, an occurrence seen in severe rickets, was present. A large lytic 6 x 2.5 cm lesion with sclerotic borders was noted at mid-femur on the right.

Following treatment with calcitriol (1.5  $\mu\text{g}/\text{day}$ ) and NeutraPhos, a phosphorus replacement

supplement (0.5 g three times daily), for 4 months the rickets improved. After discontinuing treatment for 2 weeks the lytic lesion was surgically removed. Within 15 days postoperation, serum phosphorus rose to normal levels. The patient was cured as evaluated by chemical analysis of serum and urine. The histopathology of the tumor was consistent with a diagnosis of hemangiopericytoma.

Only six other cases of HR associated with bone tumors in children have been reported in the literature. The tumors were classified as fibrous dysplasia, fibroma, osteoblastoma-like variants, and non-osseous soft tissue tumors. In adults, HR occurs with connective tissue tumors located in soft tissues that have morphologic features of hemangiopericytoma.

The authors conclude that the tumor produced a phosphaturic substance that impaired phosphate resorption by kidney tubule cells, although production of a substance inhibiting vitamin D metabo-

lism has been implicated in other cases. In this patient, an associated amino aciduria was of diagnostic import in distinguishing genetic from tumor-caused etiology. Most importantly, the authors urge that the possibility of a tumor be considered in sporadic cases of HR.

Hanukoglu A, Chalew SA, Sun CJ, et al. *Clin Pediatr* 1989;28:321-325.

**Editor's comment**—*The diagnosis of tumor could readily be missed in sporadic cases of HR. The presence of amino aciduria is found in vitamin D deficiency and vitamin D dependency rickets but not in HR unless a tumor is present. Although most physicians probably do not check for amino aciduria, the presence of this substance should be evaluated in all sporadic cases. If found, screening for tumor should follow. The absence of amino aciduria may not absolutely exclude the possibility of tumor, but it makes it much less likely.*

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