

PARATHYROID ADENOMA IN A CHILD AND HUNGRY BONE SYNDROME FOLLOWING SURGERY

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ABSTRACT

Primary hyperparathyroidism is rare in adults, but extremely rare in children under the age of 14 years. Childhood hyperparathyroidism usually becomes manifest after 10 years of age and is most frequently caused by a single adenoma. Rarely, the "hungry bone syndrome" develops following surgery. In this case report we would like to present an 11 9/12 year old girl with bilateral genu valgum deformity owing to a single parathyroid adenoma. Furthermore, she developed the "hungry bone syndrome" following parathyroid surgery.

Key Words: Hyperparathyroidism, children, hungry bone syndrome

INTRODUCTION

Primary hyperparathyroidism is almost caused by generalized hyperplasia of the parathyroid glands in the neonatal period; but onset during childhood is usually the result of a single benign adenoma. Hypercalcemia may cause muscular

weakness, anorexia, vomiting, constipation, polydipsia, polyuria, weight loss and fever. Osseous changes may produce severe pain in the back or extremities, gait disturbances, genu valgum and fractures. Prognosis is good if the disease is diagnosed early and appropriate surgical treatment is received.

CASE REPORT

An 11 9/12 year old girl presented with constipation, difficulty in climbing the stairs and bilateral genu valgum deformity. She was the fourth child of a healthy couple, both 35 years of age. She was delivered by normal spontaneous route. She had no health problem until the age of 6 years. Her family history was normal.

On physical examination; her weight was 20kg. (<3 per.), height was 118 cm. (<3 per.). A mass, 0.5 cm in diameter, was palpable in the thyroid lodge. 1-2/6 systolic murmur was heard at the apex on cardiac auscultation. She had mild pectus carinatum, rachitic rosaries, dilatation of metaphyses, genu valgum and coxa vara deformities and mild flexion contracture of hip

and knee joints. There was no other pathologic finding on the systemic examination. She was a prepubertal female.

Laboratory investigations showed hypercalcemia (Ca:11.8 mg/dl), hypophosphatemia (P:2.6 mg/dl) with elevated serum alkaline phosphatase (ALP:3042 IU/L) and parathyroid hormone levels (PTH:↑2000 pg/ml) (N.9-55 pg/ml). Thyroid hormone levels were normal. Urinary calcium/creatinine ratio was increased (Ca/Crea:3) and urinary calcium excretion was 13.4 mg/kg/day. Her skeletal survey showed generalized decreased density of bones. Roentgenograms revealed subchondral resorption on the distal ends of the clavicles and subperiosteal resorption on the proximal ends of the humerus and along the radial sides of the phalanges. Distal metaphyses of humerus and ulna and the proximal metaphyses of tibia were dilated and irregular. The inclination angle of the femur was declined, leading to coxa vara deformity. Cranial roentgenogram revealed

granular deossification on especially frontal and occipital bones. All these osseous changes were reported to be related with the primary hyperparathyroidism. On cervical ultrasonography, a hypoechoic mass lesion was visualized on the inferoposterior aspect of the right side of the neck, suggesting a parathyroid adenoma.

The parathyroid scintigraphy showed a single, 2x2 cm, parathyroid adenoma. Abdominal and urinary system ultrasonography showed no pathology. On echocardiography, minimal mitral and tricuspid regurgitation was found. Ophthalmoscopic examination revealed glaucoma. As no other pathology was found, her growth retardation was accepted to be related with hyperparathyroidism.

The results of family screening for calcium, phosphorus and alkaline phosphatase were normal. The patient was referred to a pediatric surgery unit for the removal of parathyroid

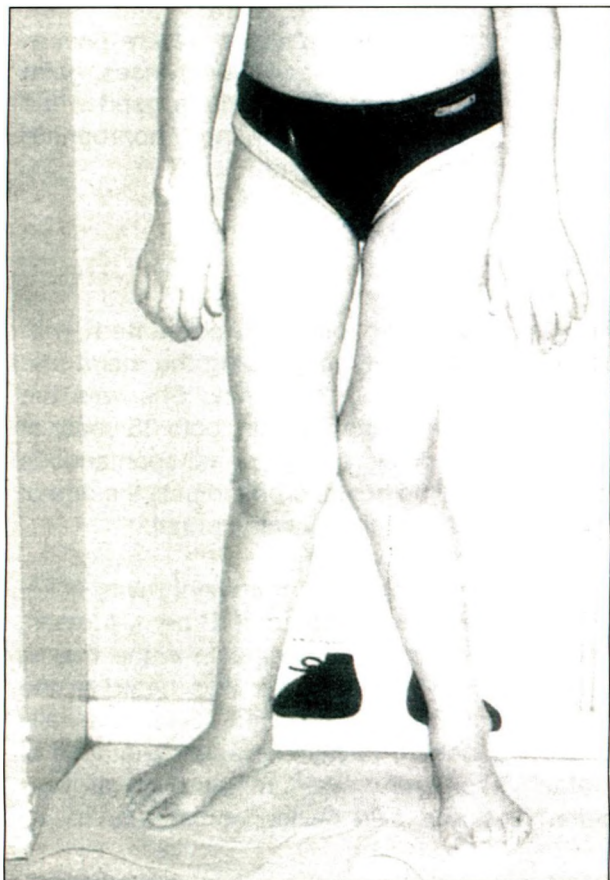


Fig.1



Fig.2

Bilateral genu valgum deformity owing to parathroid adenoma

adenoma. Histopathological examination confirmed our diagnosis of parathyroid surgery, suggesting the hungry bone syndrome. Calcium, phosphorus and active vitamin D supplementation were given and biochemical findings returned to normal in about one month. On the 26th day of the postoperative period, the serum parathyroid hormone level was 1.2 ng/ml (0.4-1.4) and the bone demineralization disappeared.

Her genu valgum deformity was successfully corrected by surgery 6 months after the parathyroidectomy.

DISCUSSION

Primary hyperparathyroidism is most common in adults after the fifth decade of life, but is very rare in children under the age of 14 years (1). Childhood hyperparathyroidism usually becomes manifest after 10 years of age and is more frequent in boys than in girls. It is most frequently caused by a single parathyroid adenoma, very few instances of parathyroid carcinoma are known in children (2). In autosomal dominant cases of hyperparathyroidism, children have been involved in about a third of the pedigrees (3). Our case was an 11 9/12 year old girl with no family history of hyperparathyroidism.

The clinical manifestations are usually subtle and that is why this disorder is usually diagnosed and treated late. Szewczyk presented the case of a 17-year old girl with parathyroid adenoma, who had been "observed" for over 12 months because of persistent ostealgia (4). Makhdoomi et al presented another case of parathyroid adenoma in a girl whose hypercalcemia was initially labeled as 'idiopathic' and therefore remained undiagnosed for 11 years in spite of repeated hospital admissions (1). Schmidt et al reviewed 71 cases with asymptomatic, persistent hypercalcemia and found solitary benign parathyroid adenoma in 62 (%88) cases, parathyroid four-gland hyperplasia in 5 (%6) cases, multiple adenoma in 2 (%3) cases and no cause in 2 (%3) cases (5). Therefore, screening of the parathyroid glands is crucial in hypercalcemia workup.

Osseous changes owing to hypercalcemia are more common in children than in adults and may

lead to pain in the back or extremities, gait disturbances, genu valgum, fractures (2,3). These osseous complications may rarely be the first symptom of hyperparathyroidism (6), as in our case, where the patient presented with bilateral genu valgum deformity. Renal calculi may produce renal colic and hematuria (3). Our case had no renal complications.

In the treatment, surgical exploration is indicated in all instances. If parathyroid adenoma is diagnosed, it should be removed. Rarely, the "hungry bone syndrome" develops in a few days following parathyroid surgery, as in our case. Since both serum calcium and phosphorus levels are decreased; calcium, phosphorus and Vit D supplementation are required in the management of this syndrome (7). According to the literature we reviewed, there is no new treatment modality for parathyroid adenoma and for its preoperative or postoperative complications.

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