Int J Endocrinol Metab 2006; 3: 176-179

CASE REPORT

Primary Hyperparathyroidism in an Adolescent Girl

Rashaud A, Najafizadeh M, Ghoraian MA

Institute of Endocrinology and Metabolism, Iran University of Medical Sciences, Tehran, I.R.Iran

The aim of this paper is to present the case of a 15.5 year old girl with primary hyperparathyroidism (PHPT) with symptoms of bone pain that appeard 2-3 years prior to admission; she had suffered bone fracture six months earlier and had high serum calcium, very high serum PTH and low serum phosphorous levels, all results being in agreement with PHPT.
The diagnosis was confirmed by imaging studies with 99m TC-Sestamibi scan, showing an adenoma in parathyroid tissue. The adenoma was removed by surgical operation. After surgery the patient was treated by high doses of calcium and vitamin D to avoid postoperative hypocalcemia.

Key Words: Primary hyperparathyroidism, Hypercalcemia

Received: 19/12/2005 - **Accepted**: 12/08/2006

Introduction

Primary Hyperparathyroidism (PHPT) is a generalized disorder of calcium, phosphate and bone metabolism that results from increased secretion of the parathyroid hormone; it most commonly occurs in adults especially the elderly. However primary hyperparathyroidism does occasionally occur in children.[1,2] PHPT may go undetected for years because of the minimal symptoms of the disease.[2] The infrequency of screening lab tests in pediatric patients' laboratory studies, including calcium and PTH levels, results in significant delays that often occur between onset of the symptoms and the time of diagnosis of primary hyperparathyroidism. Early diagnosis of the disease can help avoid metabolic consequences of hypercalcemia like hypercalcuria, nephrolithiasis, nephrocalcinosis, CNS damages, as well as bone fractures and other complications[2]. In a study conducted by Kollars et al[3] on 52 patients who underwent parathyroid resection between 1970 and 2000, the most common presenting symptoms reported were fatigue and lethargy, headache, nephrolithiasis, nausea, abdominal pain, vomiting and polydipsia.[3] An adenoma of one or more parathyroid glands is the most common cause of the disease.[4] It has been suggested that bone disease is more common whereas renal disease is less common in children.[5,6] The classical presentation of PHPT with nephrolithiasis, cystic bone disease and soft tissue calcification has become less common and has been replaced increasingly by patients presenting with mild hypercalcemia and few or no symptoms.[7]

Correspondence: A. Rashaud, Institute of Endocrinology and Metabolism, Firoozgar Alley, Valadi St., Valie-Asr Ave, Tehran, I.R.Iran.
E-mail:meh_naj2000@yahoo.com

Case Report

A 15.5 year old girl was admitted to the Institute of Endocrinology and Metabolism for evaluation of short stature. Her menstruation

onset was three years prior to admission. Height was 147cm and weight 34 kg. On admission the patient had genovalgus and rather severe pain in her knees, that had begun about 2-3 years prior to admission. About 6 months before admission there was a fracture on the femur just above the knee following mild trauma (pathologic fracture). On physical examination diffuse bilateral goiter was noticed. There was a nodule under the lower pole of the right lobe of the thyroid, dimensions approx 2x2x2cm. Laboratory examinations revealed the following results: T_3: 1.8ng/mL (normal:0.8-2.0), T_4: 4.9 µg/dL, (normal: 4.5-12), T_3 Resin-uptake: 25% (normal:25-35%), TSH, IRMA: 1.2 mIU/L (normal: 0.3-4.0).

Results for evaluation of parathyroid function showed high serum and 24-hour urine Ca levels, low inorganic phosphorous, high alkaline phosphatase activity, and alkaline pH of all urine samples. The most important finding was extremely high serum PTH levels; (Table 1). Radioisotope scan of thyroid gland with Technesium 99^m reported decreased iodine uptake by thyroid lobes and increased background activity with a non functioning nodule. Radiography of carpal and metacarpal bones, revealed osteoporosis and preosteal resorption of these bones, especially on the radial part, suggesting hyperparathyroidism (Fig. 1). According to the MIBI parathyroid scan report following injection of TC-99 MIBI, early images showed a focus of tracer uptake in the lower portion of the right thyroid lobe. On delayed images, an area of increased uptake adjacent to the lower pole of the right thyroid lobe was noticed, a finding in favor of abnormal functioning tissue (parathyroid adenoma) (Fig. 2).

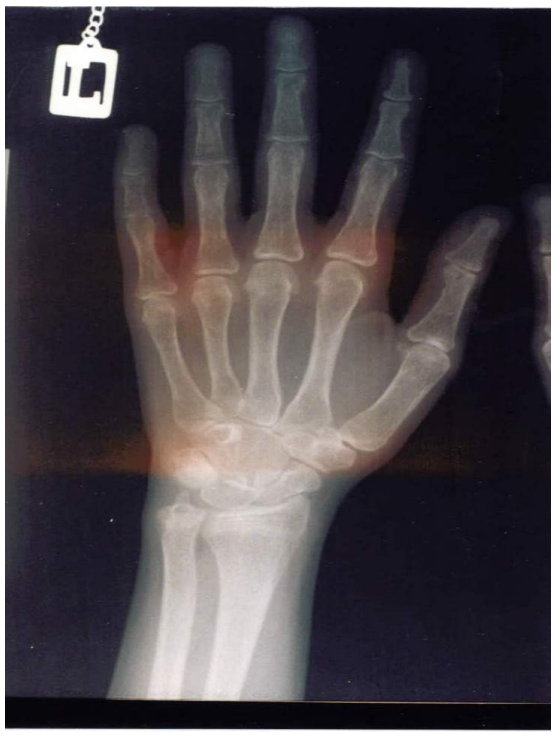

Fig.1. Radiography of hand

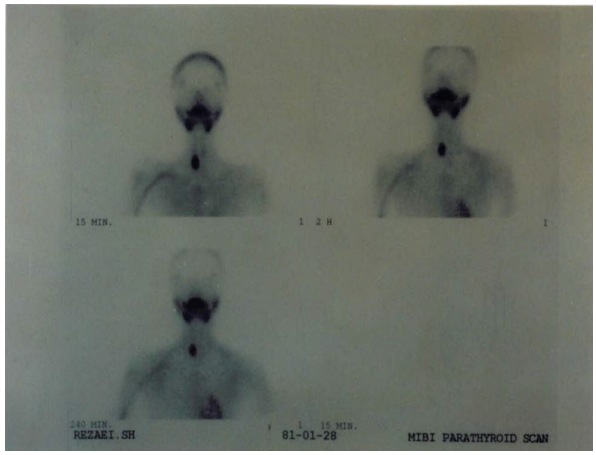

Fig.2. MIBI Parathyroid scan

Surgical operation was performed for dissection of the nodule (right lower parathyroid

Table 1. Laboratory findings prior to surgical and after operation

Variable	Before	After	Reference range
Serum Ca (mg/dL)	12.4	9.5	8.5-10.5
Serum Phosphorus (mg/dL)	2.7	3.2	Child.: 4.5-6.5 Adults: 2.5-5.0
Serum alkaline phosphatase (IU/L)	1555	-	Up to 444
24hr urine Ca (mg/24hr)	432	65	100-300
24 hr urine phosphorous (mg/24hr)	600	189	400-1000
Serum iPTH (ng/mL)	2048	51	13-54
24 hr urine creatinine (g/24hr)	0.6	0.5	0.6-1.8

ectomy). The dimensions of the adenoma were 1x 2 cm and its weight was 2 grams. The pathology report confirmed parathyroid adenoma. After surgery, high doses of calcium and vitamin D_3 were prescribed to avoid hypocalcemia due to hungry bone syndrome. Postoperative laboratory results (2 weeks after surgery) are shown in Table 1

Discussion

Primary hyperparathyroidism is a rare but serious disease, which in most cases is diagnosed by chance. Symptoms are often minimal or the disease is asymptomatic.[8]

In the study by Kollars et al,[3] it was found that PHPT is more common in adolescents than younger children (median age was 16.8 years) and the most common presenting features reported in their study were fatigue and lethargy, headache, nephrolithiasis, nausea, abdominal pain, vomiting and polydipsia. In the present study, patient symptoms included fatigue, lethargy, abdominal pain and polydipsia. Most of the symptoms are consistent with the Kollars et al report. They found that PHPT is most commonly due to a single adenoma and, in all cases, surgery had been curative. In a retrospective analysis of 16 cases of PHPT aged 10.5-20 years, 11 (85%) had single adenomas and surgery was curative in all cases. In a similar study 80% of patients with PHPT had parathyroid adenoma.[9,10]

All the above mentioned findings are consistent with our report. Hyperparathyroid syndromes including multiple endocrine neoplasia (MEN) type I and type IIA, familial hyperparathyroidism and familial cystic parathyroid adenomatosis may be seen in children.[4] When familial PHPT is diagnosed in a child, other members of the family should be screened for MEN types I and IIA.

In assessing children with hypercalcemia, it is important to exclude familial hypocalcuric hypercalcemia, which may occasionally cause elevated PTH levels. This syndrome has a benign course and parathyroid surgery is not recommended for these patients.[11] Considering elevated 24 urine calcium levels, the patient in present study did not qualify for this category.

According to the recommendations of the panel of world experts on issues related to PHPT, at a 2 day conference, the value for the total calcium concentration to be regarded as a criterion for surgery was set at 1mg/dL (0.25mM) above the upper limits of normal;[12] it was also suggested that a number of possible risk factors be considered for development of complications of primary hyperparathyroidism including:

-Serum calcium concentrations of 1 mg/dL above accepted normal range.

-Confirmed 24hour urinary calcium of more than 400mg.

-Patients under 50 years of age and

-Bone density at the lumbar spine, hip, or distal radius of more than 2.5SD below peak bone mass.

Surgery was recognized as the only definitive therapy for primary hyperparathyroidism and was acknowledged to be virtually an appropriate course of action. Accurate localization of adenoma prior to surgery is necessary for a successful operation. Using 99m Tc-sestamibi scintigraphy, a sensitive and specific parathyroid scan may be performed, resulting in reduction of operation time and morbidity.[13,14] This technique was successfully used for localization of the adenoma in our patient.

Postoperative hypocalcemia may be caused by permanent or transient hypoparathyroidism or extensive skeletal remineralisation[15] and should be treated with high doses of calcium and vitamin D.

References

1. Schmidt H, Kusser B, Spelsberg F. Asymptomatic hypercalcemia due to an ectopic parathyroid adenoma in an 8-year-old boy. Exp Clin Endocrinol Diabetes 2001; 109: 184-6.
2. Damiani D, Aguiar CH, Bueno VS, Montenegro FL, Koch VH, Cocozza AM, et al. Primary hyperparathyroidism in children: patient report and review of the literature. J Pediatr Endocrinol Metab 1998; 11: 83-6.
3. Kollars J, Zarroug AE, van Heerden J, Lteif A, Stavlo P, Suarez L, et al. Primary hyperparathyroidism in pediatric patients. Pediatrics 2005; 115: 974-80.
4. Root AW, Diamond FB Jr. Disorders of calcium metabolism in the child and adolescent. In: Sperling MA, editors. Pediatric Endocrinology. 2nd ed. Philadelphia: Saunders 2002. p. 638.
5. Girard RM, Belanger A, Hazel B. Primary hyperparathyroidism in children. Can J Surg 1982; 25: 11-3.
6. Allo M, Thompson NW, Harness JK, Nishiyama RH. Primary hyperparathyroidism in children, adolescents, and young adults. World J Surg 1982; 6: 771-6.
7. Uden P, Chan A, Duh QY, Siperstein A, Clark OH. Primary hyperparathyroidism in younger and older patients: symptoms and outcome of surgery. World J Surg 1992; 16: 791-7
8. Mollerup CL, Bollerslev J, Blichert-Toft M. Primary hyperparathyroidism: incidence and clinical and biochemical characteristics. A demographic study. Eur J Surg 1994; 160: 485-9.
9. Hsu SC, Levine MA. Primary hyperparathyroidism in children and adolescents: the Johns Hopkins Children's Center experience 1984-2001. J Bone Miner Res 2002; 17 Suppl 2: N44-50.
10. Lawson ML, Miller SF, Ellis G, Filler RM, Kooh SW. Primary hyperparathyroidism in a paediatric hospital. QJM 1996; 89: 921-32.
11. Bornemann M. Management of primary hyperparathyroidism in children. South Med J 1998; 91: 475-6.
12. Bilezikian JP, Potts JT Jr, Fuleihan Gel-H, Kleerekoper M, Neer R, Peacock M, et al. Summary statement from a workshop on asymptomatic primary hyperparathyroidism: a perspective for the 21st century. J Clin Endocrinol Metab 2002; 87: 5353-61.
13. Lundgren EC, Gillott AR, Wiseman JS, Beck J. The role of preoperative localization in primary hyperparathyroidism. Am Surg 1995; 61: 393-6.
14. Worsey MJ, Carty SE, Watson CG. Success of unilateral neck exploration for sporadic primary hyperparathyroidism. Surgery 1993; 114: 1024-9
15. Brasier AR, Nussbaum SR. Hungry bone syndrome: clinical and biochemical predictors of its occurrence after parathyroid surgery. Am J Med 1988; 84: 654-60.