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Short Communication

Boerhaave's Syndrome: an Unusual Presentation of A Pediatric Giant Parathyroid Adenoma

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Abstract

This is a case of a 15 year old, otherwise healthy, girl who presented with severe abdominal pain, nausea and vomiting. She was found to have acute pancreatitis and esophageal perforation "Boerhaave's syndrome" as a complication of elevated serum calcium level of 15.3 (reference range, 9.3 mg/dL-10.6 mg/dL). Neck ultrasound showed 2.7 cm mass in the right side posterior to the right thyroid lobe. During surgical neck exploration, an enlarged parathyroid adenoma was found. After surgery, she was eucalcemic, and recovered from pancreatitis and Boerhaave's syndrome with no subsequent issues. In the month following surgery, her serum calcium level was 9.1 and her parathyroid hormone level was 80.

Keywords: Multiple endocrine neoplasia; Parathyroid hormone; Primary hyperparathyroidism

Introduction

A previously healthy 15-year-old girl presented to a local emergency department with severe abdominal pain and vomiting. She had constipation for 10 days prior to that. She had a long standing abdominal pain and constipation, which were diagnosed as functional, apart from that, she had no significant past medical, surgical or family history. In the emergency department, she was tachycardiac to 110s, and normotensive. Her abdomen was tender in the epigastric area with guarding. Blood work showed white blood cell count of 18,600/µL; hemoglobin, 12.7 g/dL; total serum calcium 15.3 mg/dl; ionized calcium 7.9; and pancreatic lipase 573 U/L (Table 1). CT chest, abdomen and pelvic showed peripancreatic fluid collection and pneumomediastinum extending through the mediastinum to the distal esophagus without pneumothorax. She was diagnosed with acute pancreatitis secondary to hypercalcemia and Boerhaave's syndrome secondary to severe retching and vomiting. On the workup for her hypercalcemia, her Parathyroid Hormone (PTH) level was 897 pg/ml (reference range, 15 pg/ml-65 pg/ml). She had neck ultrasound which showed 2.7 cm right parathyroid gland (Figure 1).

Management

After fluid resuscitation and stabilization of the patient, surgery was done. Markedly enlarged right superior parathyroid gland was noted and resected (Figure 2). Intraoperative parathyroid hormone level decreased appropriately from 1605 pg/ml to 45 pg/ml after 30 minutes of gland removal. She was treated conservatively for the pancreatitis and Boerhaave's syndrome.

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Post operatively, the patient developed mild symptoms of hypocalcemia, which were due to impaired bone mineralization while hyperparathyroid compounded by dropping of serum calcium level from very high to normal. Her serum calcium was ranging between 7.8 mg/dL to 8.7 mg/dL. She was treated with calcium and vitamin D supplementation, she required 74 mg/kg/day of elemental calcium to maintain a normal level of her serum calcium. Her condition continued to improve postoperatively, and she was discharged on calcium 1000 mg, 4 times a day, calcitriol 1 mcg twice a day, and 5000 units of vitamin D supplements. On follow up visit after 1 month, her serum calcium was 9.1 mg/dL and her PTH level was 83 pg/mL. She was scheduled for outpatient Medical Genetics to test for hyperparathyroidism-related syndromes.

Discussion

Primary Hyperparathyroidism (PHPT) occurs primarily due to increased autonomous production of Parathyroid Hormone (PTH) from the parathyroid glands without stimulating low serum calcium levels. The main pathologic conditions in PHPT are adenoma, glandular hyperplasia and carcinoma. Single parathyroid gland adenoma accounts for 80% to 85% of PHPT [1]. Parathyroid adenomas consist mainly of parathyroid chief cells; however, some are composed of oxiphil cells which usually tend to be larger sized adenomas compared to those of chief cells.

Table 1: Preoperative and postoperatvive lab values of the patient and reference ranges.

Blood	preoperative	postoperative	Reference range
WBC	18,600/µL	5.1	3800-10400
HB	12.7 g/dl	8.1	11.9-14.8
Calcium	15.3 mg/dl	10.2	9.3-10.6
Calcium, ionized	7.9 mg/dl	-	4.9-5.5
Pancreatic lipase	573 U/L	199	13-60
PTH	897 pg/ml	22	15-65
Sodium	135 mmol/l	142	135-145
Potassium	2.9 mmol/l	3.7	3.6-5.2
BUN	33 mg/dl	21	20-Jul
Creatinine	1.52 mg/dl	1.53	0.35-0.86
Magnesium	1.5 mg/dl	1.6	1.6-2.3
Phosphorus	2.4 mg/dl	2.8	3.5-4.9

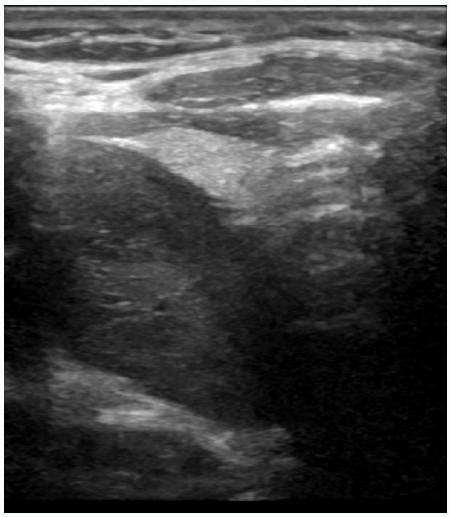

Figure 1: Neck US showing enlarged right parathyroid gland.

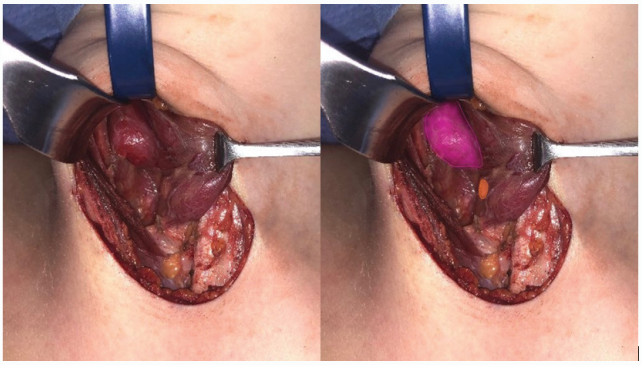

Figure 2: Intraoperative view of right superior (highlighted in pink) and inferior (orange) parathyroid glands.

Causes of PHPT are single gland adenoma or multiple gland hyperplasia. Solitary adenoma is the most common cause of primary hyperparathyroidism in children [2]. PHPT can be due to isolated sporadic cases of parathyroid adenoma or part of Multiple Endocrine Neoplasia (MEN) I or II or familial non-MEN HPT as a result of multigland parathyroid hyperplasia.

PHPT occurs primarily in adults and it is rare in children [2,3], with incidence of 2 to 5 per 100,000 [2]. Most children present with nonspecific symptoms such as polyuria, fatigue, poor appetite, weight loss, abdominal pain, nausea, and emesis [2]. Due to its rare occurrence and the non-specific symptoms, the diagnosis is often delayed, and it is usually after development of secondary complications. In our patient, hyperparathyroidism wasn't diagnosed prior to the onset of acute complication of hypercalcemia. Although she was having constipation for a period of time prior to the onset, calcium and PTH levels weren't checked as constipation being nonspecific symptom with multiple variant etiologies.

The diagnosis of PHPT in children is primarily by biochemical serum levels of calcium and parathyroid hormones, and by imaging localizing the parathyroid gland such as ultrasound, CT or sestamibi scans. Neck US was used in the diagnosis in our case. Surgical resection is the treatment of choice for HPT in children [2,4].

Hypercalcemia is one of the commonest electrolyte derangements that occur in hyperparathyroidism. Acute pancreatitis has been considered as a complication of primary hyperparathyroidism. The prevalence of acute pancreatitis in PHPT has been estimated to be between 1.5% and 13% [5]. There are different mechanisms that explain the development of acute pancreatitis in PHPT. Hypercalcemia from PHPT leads to de novo activation of trypsinogen to trypsin, resulting in autodigestion of the pancreas and subsequent pancreatitis. Hypercalcemia also leads to formation of pancreatic calculi which lead to ductal obstruction and pancreatitis [5]. High calcium level in our patient (15.3 mg/dL) was suspected to be the triggering event in the development of acute pancreatitis.

The principles of management of pancreatitis in children are similar to those in adults. Fluid resuscitation is crucial to replace third spacing that occurs acutely with the severe cases. Adequate nutritional support is also important. Our patient needed to be on peripheral parenteral nutrition for a short period of time to ensure adequate nutritional replacement, and as part of the conservative management of her esophageal perforation. Pain control is also necessary. Antibiotics are generally not needed, even in the presence of elevated white blood cell counts and fever; they should be used only when infection is strongly suspected [6]. Surgical management might only be needed in cases of duodenal or biliary obstructing lesions, or in children with recurrent pancreatitis due to intra-pancreatic ductal strictures.

Boerhaave's syndrome is rare in children less than 18 years, with only limited cases reported in the literature [7]. Although in adults it occurs spontaneously after vomiting, in children it is more often due trauma or iatrogenic. Increased intraluminal pressure leads to perforation in the lower part of the esophagus. In most children, the rupture in the distal esophagus is on the right side (into the right pleural cavity), in contrast to adults, where usually a left-sided rupture is found [7]. The diagnosis can be confirmed by the findings of pneumomediastinum in CXR or CT scan after bouts of retching and vomiting. In our patient, the CT scan of the chest showed pneumomediastinum extending throughout the mediastinum and into the base of the neck with distal involvement extending to the distal esophagus. Conservative management in the treatment of Boerhaave's syndrome can be applied to patients with a small defect, with contamination limited to the mediastinum, and to patients with a late diagnosis (>24 hours delay) [8]. However, some reviews showed surgery being the treatment of choice, with primary closure of the rupture, partial resection of the esophagus, drainage, or an intraluminal stent [7]. In our patient, since the perforation was small, without complications related to it, it was decided to treat it conservatively, by keeping the patient NPO, peripheral nutrition, adequate IV hydration and antibiotics.

Conclusion

Primary hyperparathyroidism causing malignant hypercalcemiais rare, and pancreatitis and Boerhaave's syndrome are very unusual presentation in children. Parathyroid adenomas are usually well circumscribed and easy to excise. Pancreatitis and Boerhaave's syndrome usually managed conservatively and should not recur once the calcium level normalizes after excision of adenoma.

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