Parathyroid crisis in a 20 year old—an unusual cause of hypercalcaemic crisis

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Abstract
Since the advent of automated serum analysis, patients with primary hyperparathyroidism (PHPT) are often asymptomatic at presentation or have mild symptoms attributable to the disease. Parathyroid crisis is a rare and potentially fatal complication of PHPT in which patients develop severe hypercalcaemia with signs and symptoms of multiple organ dysfunction. A case of parathyroid crisis in a 20 year old man who presented with brown tumours and renal stones is described.

Case report
A 20 year old college student presented with left knee pain after a minor sport’s injury. Plain radiographs of the knee demonstrated multiple lytic bone lesions in the distal femur and proximal tibia, which were confirmed by magnetic resonance imaging. Bone scan demonstrated generalized increased uptake with focal moderate activity around the left knee. High-resolution ultrasound demonstrated a parathyroid adenoma (arrows) inferior to the left lobe of the thyroid, feeding parathyroid adenoma.

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469

Hypercalcaemic crisis is characterised by severe hypercalcaemia, usually with a calcium >3.5 mmol/l (>14 mg/dl), with associated signs and symptoms involving multiple organ systems. Patients often display metabolic encephalopathy, renal insufficiency, gastrointestinal symptoms, and cardiac dysrhythmia. Malignancy is the most common cause of hypercalcaemic crisis, which results from secretion of PTH related polypeptide or from bone resorption related to osseous metastases. Cancers most commonly associated with hypercalcaemia include squamous cell carcinoma of the lung, breast cancer, renal cell carcinoma, bladder cancer, and multiple myeloma.

Hypercalcaemia occurring with a raised intact PTH is pathognomonic for primary hyperparathyroidism (PHPT). Severe hypercalcaemia occurs in 1%–2% of patients with PHPT and has been referred to as acute hyperparathyroidism, parathyroid crisis, parathyroid storm, and parathyrotoxicosis. Other complications which are common in patients with parathyroid crisis include kidney stones, renal insufficiency, brown tumours, proximal muscle weakness, neuropsychiatric syndrome with decreased ability to concentrate and depression, and a normochromic, normocytic anaemia. Hypercalcaemic crisis has been reported to be as high as 50% in children age

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18 or younger. It is unclear whether the higher rate of crisis in the paediatric population is due to delayed diagnosis or a more aggressive form of the disease. The occurrence of hyperparathyroidism at an early age, as in this patient, should also raise the suspicion of multiple endocrine neoplasia type 1.

The initial treatment of hypercalcaemic crisis is the same regardless of the aetiology. Therapy is aimed at increasing renal excretion of calcium and decreasing bone resorption. Patients with hypercalcaemic crisis are volume depleted and initial therapy should be intravenous hydration with isotonic saline. The conventional use of loop diuretics to promote calciuresis is currently under scrutiny as this may exacerbate extracellular volume depletion through concomitant sodium diuresis. In patients with renal failure, dialysis with a low or zero calcium dialysate should be initiated. Bisphosphonates, which directly inhibit osteoclast activity, are the most effective of the pharmacological agents that decrease bone resorption. Pamidronate has gained acceptance as the drug of choice since it is the most potent bisphosphonate and has the fewest side effects.

After initial medical stabilisation, surgical exploration should be performed on patients with parathyroid crisis. Recent series have reported excellent results from urgent parathyroidectomy, with perioperative mortality of less than 5%. As for all patients with PHPT, approximately 85%–90% of patients with parathyroid crisis have a single adenoma. As exemplified in our patient, serum PTH levels decline rapidly after parathyroidectomy, while serum calcium normalises over the course of several days. The correction of serum calcium and PTH is accompanied by resolution of bone pain, renal insufficiency, cardiac dysrhythmias, and mental status changes. Complete resolution of the metabolic derangements including resolution of bone, renal, and haematological abnormalities may take six to 12 months.

Conclusion
Hyperparathyroidism must be considered in the differential diagnosis for severe hypercalcaemia in all age groups. Patients can present with a wide range of symptoms related to metabolic derangements of multiple organ systems. Initial medical management is aimed at lowering serum calcium by increasing renal excretion of calcium and reducing bone resorption. Patients should be treated by urgent parathyroidectomy. Surgical treatment results in prompt correction of serum calcium and PTH with gradual resolution of the metabolic consequences of severe hypercalcaemia.

Learning points

- Primary hyperparathyroidism (PHPT) must be considered in the differential diagnosis for hypercalcaemic crisis.
- Hypercalcaemia in the setting of a raised intact parathyroid hormone is pathognomonic for PHPT.
- Although PHPT is rare in young adults and children, they often present with hypercalcaemic crisis.
- Parathyroid crisis requires immediate medical management for hypercalcaemia followed by urgent surgery.
- Normalisation of serum calcium concentrations leads to gradual reversal of metabolic abnormalities associated with hypercalcaemia.

References


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