

Idiopathic hypoparathyroidism: A case study

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Abstract

Background and objective: Hypoparathyroidism is an uncommon endocrine-deficiency disease characterized by low serum calcium levels, elevated serum phosphorus levels, and absent or inappropriately low levels of parathyroid hormone in the circulation. It is characterized by carpal and pedal muscle spasms, positive Chvostek's and Trousseau's signs. Diagnosis is done by typical clinical and laboratory findings. Current treatment options include oral calcium, vitamin D, and thiazide diuretics. We present a 21 years old girl exhibiting characteristic features of idiopathic hypoparathyroidism presented with non-union fracture of right ulner bone.

Keywords: Endocrine system diseases, Parathyroid hormone.

Introduction

Chronic hypocalcemia is commonly due to inadequate levels of parathyroid hormone¹. Hypoparathyroidism is a disorder of parathyroid hormone deficiency caused by two main etiologies. Autoimmune destruction of the parathyroid glands can occur as an isolated endocrine deficiency syndrome or in connection with failure of other endocrine glands. The other common etiology of hypoparathyroidism is after neck surgery in which all parathyroid tissue is removed, either in the context of surgery for primary hyperparathyroidism or after extensive neck surgery for thyroid cancer². Important physical signs of hypocalcemia are chvostik's and trousseau's signs which can be elicited in symptomatic patients with hypoparathyroidism³. Trousseau sign is a more reliable sign present in 94% of hypocalcemic individuals and only 1% to 4% of healthy people. It is the presence of carpopedal spasm observed following application of an inflated blood pressure cuff over systolic pressure for three minutes in hypocalcemic patients¹. Other clinical features of hypoparathyroidism include circumoral numbness, paresthesias, laryngeal spasm, tetany and/or seizures². Rare clinical features have been

recorded in patients with hypoparathyroidism like reversible congestive heart failure⁴, stable myelofibrosis⁵, and spontaneous intracerebral bleeding in the setting of chronic intracerebral calcifications⁶. Typical laboratory findings in patients with hypoparathyroidism include low or undetectable PTH (< 15 pg/mL), low calcium (< 8 mg/dL). For an accurate interpretation of total calcium, (its values should be corrected for serum albumin: for each 1 g/dL of albumin below 4 g/dL, 0.8 mg/dL should be added to total calcium measurement), and increased phosphorus (> 5.0 mg/dL)⁷. Maintenance treatment of hypoparathyroidism consists of correction of calcium levels by administration of oral calcium and synthetic 1 alphas-hydroxylated vitamin D. The administration of active forms of vitamin D is required due to the absence of parathyroid hormone which is the main stimulus for renal conversion of 25 hydroxyvitamin D into 1,25 dihydroxyvitamin D⁷.

Case history:

A 21 years old girl presented with nonunion fracture of right ulner bone. The fracture caused by a car accident 5 months before presentation. She was complaining of generalized weakness, numbness and

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parasthesia of both hands with attacks of hand stiffness. She also described circum-oral numbness, She had no history of neck surgery, radiation, infiltrative disease or any previous systemic illness. Family history of skeletal abnormality was negative as was for any similar conditions. Examination revealed a normal stature without any facial abnormalities, no dental problems, no mucous membrane complains or abnormalities. Eye examination was normal and both Chvostic's and Troussiuous signs were positive. No clinical evidence of any other endocrine dysfunction. Her baseline laboratory evaluation revealed:

Serum Albumin: 4.37 G/100 ml	reference range: (3.2-4.5 mg/100ml)
Serum Calcium: 4.8 mg/100 ml	reference range (9.2-11 mg/100ml)
Serum Phosphorus: 6.2 mg/100 ml	reference range (2.3-4.7 mg/100ml)
Serum parathyroid hormone: 9 pg/ml	reference range (9.4-81.6 Pg/ml)
24 hour urinary calcium: 27.9mg/24 hour	reference range (100-240 mg/24 hour)
Serum magnesium: 2 mg/100 ml	reference range (1.7-2.2 mg/100ml hour)

CBC:

WBC: $5.71 \times 10^3/\mu\text{L}$, Hb: 12.8 g/100ml, Platelets $230 \times 10^3/\mu\text{L}$, ESR: 12 mm/Hour

Blood urea: 26.9 mg/100ml

Serum Creatinin: 0.69 mg/100m

Serum sodium: 143 mg/100ml

Serum potassium: 3.4 meq/100ml

Electrocardiography revealed: normal sinus rhythm, one mm ST segment depression and T wave inversion in lateral chest leads while QT interval was within normal range. X ray of her right hand and forearm revealed displaced fracture of ulner bone with normal bone density. Calcium and active vitamin D started as long term treatment with follow up and orthopedic care.

Discussion

Patients presenting with features suggestive of hypocalcemia should undergo laboratory evaluation. Hypocalcemia should be confirmed by measuring serum calcium and serum albumin. If albumin level was below 4 g/100ml serum calcium is corrected accordingly. In the current patient serum albumin is more than 4g/100ml and serum calcium remain as the same which is less than normal, then serum phosphorus is measured and if its level is more than normal in the presence of low serum parathyroid hormone level is typical of hypoparathyroidism as in the current case⁷. Regarding classification of the disease, 4 types have been identified;

- ١- Idiopathic hypoparathyroidism
- ٢- Pseudohypoparathyroidism
- ٣- other hypoparathyroidism
- ٤- Procedural hypoparathyroidism

In the current patient there is no place for pseudohypoparathyroidism since parathyroid hormone level will be elevated in that condition and there will be abnormal musculoskeletal features⁸. On the other hand, absence of previous neck surgical procedures, and lack of evidence of any infiltrative or granulomatous diseases make idiopathic hypoparathyroidism most likely. Two features give some identity to this case: presentation with nonunion fracture and age at time of presentation as these patients are usually present within the first decade of life though may appear later⁸.

Conclusion

Cases of idiopathic hypoparathyroidism are present in Iraqi-Kurdistan and may present unusually.

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