CASE REPORT

Severe Isolated Primary Hypoparathyroidism in an Adult

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ABSTRACT

A man aged 42 years, presented with 3 years history of paraesthesias in hands and feet and muscle cramps off and on, progressing to severe carpopedal spasm, a couple of times, relieved by intravenous calcium gluconate at the emergency reception of the hospital. On examination, Trousseau's sign and Chvostek's sign were positive. Thyroid gland was not enlarged. Right eye showed mature cataract. Total serum calcium, corrected serum calcium, serum phosphate, ionized serum calcium, serum alkaline phosphatase, serum parathormone (PTH) level were deranged favouring hypoparathyroidism. He was diagnosed to be suffering from isolated primary hypoparathyroidism and put on alfacalcidol and oral calcium carbonate, with which he is asymptomatic now.

Key words: *Primary hypoparathyroidism. Hypocalcaemia. Carpopedal spasm. Chvostek's sign. Trousseau's sign.*

INTRODUCTION

Acquired hypoparathyroidism not related to surgery is most often an autoimmune disease.[1] Permanent hypoparathyroidism can result from immune-mediated destruction of the parathyroid glands.[1] Alternatively, hypoparathyroidism may result from activating antibodies to the calcium sensing receptor that decrease parathormone (PTH) secretion.[2,3] Autoimmune hypoparathyroidism is a common feature of polyglandular autoimmune syndrome type I, which is a familial disorder. This syndrome typically presents in childhood with candidiasis, followed several years later by hypoparathyroidism, and then adrenal insufficiency during adolescence.[3,4]

We are presenting here a case of isolated primary hypoparathyroidism in an adult who presented with severe hypocalcaemia.

CASE REPORT

A man aged 42 years, an office clerk by profession, presented with 3 years history of paraesthesias in hands and feet and muscle cramps off and on. He had carpopedal spasm about 3 years back after an episode of mild diarrhoea. The symptoms had gradually increased in the previous 3 months and he was having frequent muscle cramps, paraesthesias in hands and severe carpopedal spasm, a couple of times, relieved by intravenous calcium gluconate at the emergency reception of the hospital. A practitioner prescribed him tablet calcium carbonate 1.2 g, which he had been using two or three times a day irregularly, and used to feel better with that. There was no history of any thyroid illness or thyroid surgery. There was no history of dizziness, weight loss, chronic diarrhoea or abdominal pain and pigmentation of skin. He was non- diabetic and non-hypertensive. There was no family history of diabetes or hypertension. He had 2 brothers, 50 and 41 years old, and a sister 35 years of age. None of the siblings had suffered from hypocalcaemia/tetany, Addison's disease or any autoimmune illness. He was a non-smoker and non-alcoholic.

On clinical examination, he was an adult of average built, anxious and apprehensive. His pulse rate was 80 beats per minute, regular in rhythm, blood pressure 130/80 mmHg in recumbency, with no significant postural drop and respiratory rate of 16 per minute. His hydration was fair. There was no pigmentation of skin or skin creases, no mucocutaneous candidiasis. Trousseau's sign and Chvostek's sign were positive. Thyroid gland was not enlarged. Right eye showed mature cataract. Rest of the general physical and systemic examination was unremarkable. Serum urea was 6.7 mmol/l, sodium was 139 mmol/l, potassium was 4.2 µmol/l and creatinine was 102 µmol/l. Total serum calcium was 1.44 mmol/l (reference range: 2.1-2.65 mmol/l), serum albumin was 49 g/l, corrected serum calcium was 1.4 mmol/l, serum phosphate was 1.52 mmol/l (reference range: 0.8-1.65 mmol/l), ionized serum calcium was 1.23 mmol/l (reference range: 1.16-1.32 mmol/l), serum alkaline phosphatase was 132 U/l, serum magnesium was 0.84 mmol/l (reference range: 0.6-1.0 mmol/l and serum parathormone (PTH) level was 0.36 pmol/l (reference range: 0.8-6.0 pmol/l). Serum free thyroxin level was 10.8 pmol/l (reference range: 7.0-21.0 pmol/l), TSH was 1.2 uIU/ml (reference range: 0.4-4.5) serum cortisol at 9 a.m. was 18 ug/dl. Serum was negative for RA (rheumatoid arthritis) factor, antinuclear antibody and antithyroid antibodies. Plain radiograph of skull did

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not reveal any basal ganglia calcification. All other investigations like haematological profile, plain radiographs of chest and abdomen and ultra-sonographic examination of abdomen were unremarkable.

He was diagnosed to be suffering from isolated primary hypoparathyroidism and put on alfacalcidol and oral calcium carbonate. The dose adjusted while monitoring his serum calcium level. On alfacalcidol 1.5 µg daily and 1 g elemental calcium orally daily, he was asymptomatic with a total serum calcium level of 1.83 mmol/l and 24 hours urine calcium excretion was 214 mg.

DISCUSSION

Among the symptoms of hypocalcaemia, tetany; papilledema, and seizures may occur in patients who develop hypocalcaemia acutely.[5,6] By comparison, ectodermal and dental changes, cataracts, basal ganglia calcification, and extrapyramidal disorders are features of chronic hypocalcaemia. These last findings are most common in patients with hypoparathyroidism. Extensive bilateral cerebral calcifications have also been reported in patients of primary hypoparathyroidism.[7]

This patient presented with a fairly long history of mild symptoms of neuromuscular irritability and cataract in one eye, but had gradually developed severe hypoparathyroidism and severe hypocalcaemia, so presented with frequent episodes of tetany. Though primary hypoparathyroidism is a chronic disorder but a case like this suggests that the acute clinical signs and symptoms of severe primary hypoparathyroidism are the same as those of acute hypocalcaemia, ranging from tingling to tetany and may be intractable generalized tonic-clonic seizures.[8]

Polyglandular autoimmune syndrome type I (APS1) is a rare autosomal recessive disorder in which females are affected slightly more frequently than males.[2] Hypoparathyroidism or chronic mucocutaneous candidiasis is usually the first manifestation, characteristically appearing during childhood or early adolescence. The hypoparathyroidism may or may not occur in association with antiparathyroid gland antibodies that are directed against the calcium-sensing receptor.[3,4] Adrenal insufficiency usually develops later, at the age of 10-15 years. This patient is in the fifth decade of life and suffering from isolated primary hypoparathyroidism only.

Most patients with hypoparathyroidism require lifelong calcium and vitamin D supplementation. The goals of therapy are to relieve symptoms, to raise and maintain the serum calcium concentration in the low-normal range, e.g. 8.0-8.5 mg/dl (2.0-2.1 mmol/L), and to avoid hypercalciuria (maintain 24-hour urinary calcium below 300 mg). All patients with permanent hypoparathyroidism require adequate calcium intake (1.0-1.5 g elemental calcium daily).[9] A variety of vitamin D preparations can be used to treat hypocalcaemia in hypoparathyroidism; active forms of vitamin D3, alfacalcidol and calcitriol are preferred because of their shorter duration of action.[10] This patient became asymptomatic on alfacalcidol 1.5 µg with 1 g of elemental calcium daily and his serum calcium was 1.83 mmol/l. Recombinant PTH has also been used to treat hypoparathyroidism but is much more expensive than standard therapy with calcitriol. However, in patients with refractory hypercalciuria, PTH is a reasonable option.

REFERENCES

1. Favus MJ, editor. Primer on the metabolic bone diseases and disorders of bone metabolism.6th ed. Philadelphia: *Lippincott William & Wilkins*; 1999.
2. Eisenbarth GS, Gottlieb PA. Autoimmune polyendocrine syndromes. *N Engl J Med* 2004; **350**:2068.
3. Kifor O, McElduff A, LeBoff MS, Moore FD Jr, Butters R, Gao P, *et al.* Activating antibodies to the calcium-sensing receptor in two patients with autoimmune hypoparathyroidism. *J Clin Endocrinol Metab* 2004; **89**:548-56. Comment in: p. 544-7.
4. Soderbergh A, Myher AG, Ekwall O, Gebre-Medhin G, Hedstrand H, Landgren E, *et al.* Prevalence and clinical associations of 10 defined autoantibodies in autoimmune polyendocrine syndrome type I. *J Clin Endocrinol Metab* 2004; **89**:557-6.
5. Tohme JF, Bilezikian JP. Hypocalcemic emergencies. *Endocrinol Metab Clin North Am* 1993; **22**:363-75.
6. Cooper MS, Gittoes NJ. Diagnosis and management of hypocalcemia. *BMJ* 2008; **336**:1298.
7. Vassilatou E, Pikounis V, Economopoulos T, Hadjidakis D. Extensive bilateral cerebral calcifications in a patient with primary hypoparathyroidism. *Arch Neurol* 2010; **67**:888-9.
8. Armelisasso C, Vaccario ML, Pontecorvi A, Mazza S. Tonic-clonic seizures in a patient with primary hypoparathyroidism: a case report. *Clin EEG Neurosci* 2004; **35**:97-9.
9. Shoback D. Clinical practice: hypoparathyroidism. *N Engl J Med* 2008; **359**:391-403.
10. Halabe A, Arie R, Mimran D, Samuel R, Liberman UA. Hypoparathyroidism: a long-term follow-up experience with 1 alpha-vitamin D3 therapy. *Clin Endocrinol (Oxf)* 1994; **40**:303-7.