Tertiary hyperparathyroidism. A long term complication of pseudohypoparathyroidism type 1b?

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Introduction

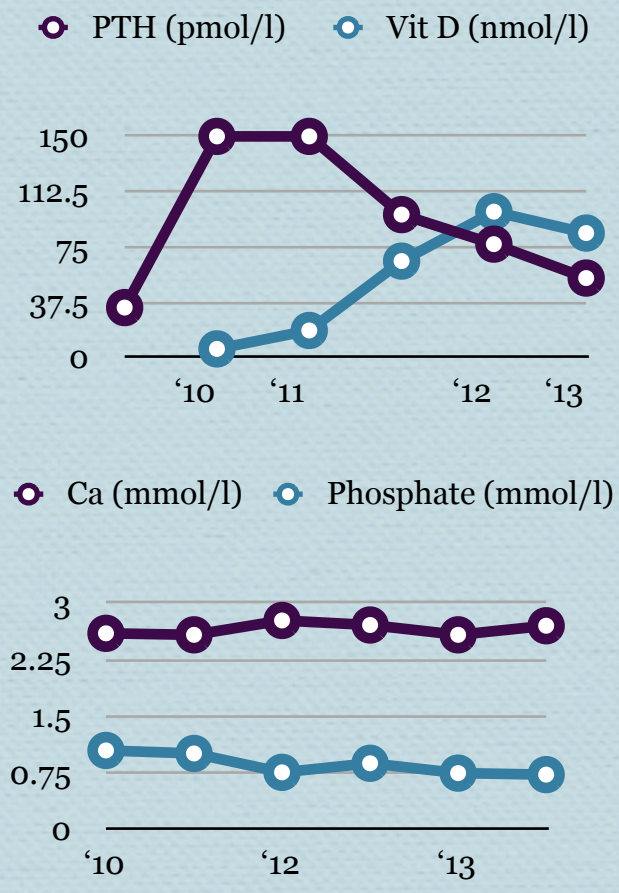
Pseudohypoparathyroidism (PHP) is a rare group of heterogenous disorders. PHP-Ib consists of renal resistance to PTH in the absence of other physical or endocrine abnormalities and is associated with reduced 1,25-OH vitamin D synthesis, increased phosphate secretion and hypocalcaemia. Despite calcium and vitamin D replacement many patients still have chronically elevated PTH.

Case description

A 31 year old white female presented with symptoms of tetany with her bloods revealing hypocalcaemia of 2.05mmol/L, increased ALP (263IU/L) and increased PTH(1100ng/L). She had normal 25-hydroxycholecalciferol. The hand radiograph suggested osteitis fibrosa. Secondary hyperparathyroidism was excluded and diagnosis of pseudohypoparathyroidism was made and patient was commenced on vit D2 and calcium supplements.

Twenty five years later she presented with hypercalcaemia, raised PTH and borderline low vit D levels and was switched from calcitriol to alpha-calcidiol which she stopped as she couldn't tolerate the preparation. Her Calcium levels remained elevated, vit D became undetectable and PTH soon exceeded 150pmol/L. The DEXA scan showed L1-L4 osteopenia and US KUB- marked bilateral cortical thinning. US neck suggested left lower parathyroid adenoma confirmed by a Sestamibi scan which revealed a well defined 2x1cm left inferior parathyroid adenoma and possibly a second intrathyroid 8mm parathyroid adenoma.

Patient was eventually established on 2200 units of cholecalciferol OD and weekly alendronate with normalisation of calcium and vitamin D levels and gradual decline in PTH levels. A decision was made not to proceed with parathyroidectomy as this could potentially lead to hyperplasia of remaining parathyroid glands.



Discussion

Chronically elevated PTH may lead to development of autonomous parathyroid tumors and tertiary hyperparathyroidism. Neary et al reported 5 patients with PHP1b who developed a parathyroid adenoma and symptomatic hypercalcaemia 21-42 years since diagnosis.

Conclusion

The management goal in PHP1b is to achieve normocalcaemia and maintain PTH as low as possible.

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Ref.: Neary et al. Development and treatment of tertiary hyperparathyroidism in patients with pseudohypoparathyroidism type 1b. JCEM.2012 Sept;97(9):3025-30