Pseudohypoparathyroidism presenting as severe Parkinsonism

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Summary
A case of pseudohypoparathyroidism in a middle-aged female presenting with severe Parkinsonism is reported. Correction of serum calcium led to marked clinical improvement suggesting that symptoms and signs were exacerbated by hypocalcaemia and tetanic spasm. Calcification of the basal ganglia was not detected on skull X-ray but was revealed by computerized axial tomography.

Introduction
In pseudohypoparathyroidism, hypocalcaemia results from receptor tissue resistance to circulating parathyroid hormone. Although calcification of the basal ganglia is associated with the condition clinical evidence of basal ganglia disease is rare. A case is reported of pseudohypoparathyroidism in a patient who presented with severe Parkinsonism.

History and examination
A 58-year-old confused, disorientated female was referred to a medical out-patient clinic because of severe tremor. According to her husband she had become increasingly withdrawn and uncommunicative over the previous 2 years. He had noted tremor in all limbs and described 4 brief episodes of total rigidity.

On examination she had classical features of Parkinsonism including immobility, mask-like facies, pill-rolling tremor, marked cogwheel rigidity and positive glabellar tap. Bilateral pes cavus was observed but no specific somatic features associated with pseudohypoparathyroidism. Trousseau's and Chvostek's signs were positive and shortly after admission she had an episode of tetany which responded to i.v. calcium gluconate.

Investigations
Biochemical measurements were as follows (normal values in parentheses): serum calcium 1.24 mmol/l (2.2-2.6 mmol/l); ionized calcium 0.5 mmol/l (1.1-2.0 mmol/l); serum inorganic phosphate 2.65 mmol/l (0.8-1.4 mmol/l); alkaline phosphatase 445 i.u./l (80-280 i.u./l); serum magnesium 0.62 mmol/l (0.7-1.0 mmol/l); parathyroid hormone 640 ng/l (undetectable to 600 ng/l). Normal laboratory studies included ESR, haemoglobin, serum sodium, potassium, chloride, bicarbonate, urea, liver function tests, plasma proteins, VDRL, 72-hr faecal fat, and thyroid function tests. Radiological skeletal survey showed generalized osteopenia with compression fractures of several vertebrae. Bone scintigraphy with 99mTc-hydroxy-ethylidene diphosphonate (HEDP) showed the vertebral fractures and also multiple rib fractures. Whole-body retention of 99mTc-HEDP measured by the method of Fogelman et al. (1980) was elevated at 40% (normal 26%) suggestive of increased bone turnover. Iliac crest biopsy confirmed increased bone resorption with no evidence of osteomalacia. No basal ganglia calcification was seen on skull X-ray but was revealed by CAT (Fig. 1).

Treatment with i.v. calcium gluconate and oral 1 α-hydroxycholecalciferol returned serum calcium and magnesium to normal with considerable clinical benefit. Rigidity became much less marked and Parkinsonian features improved. The acute effects of i.v. parathyroid hormone were measured according to the method of Tomlinson, Hendry and O'Riordan (1976). The lack of response of plasma and urine adenosine 3', 5'-cyclic monophosphate (cAMP) to bovine parathyroid hormone led to a diagnosis of pseudohypoparathyroidism type I (Fig. 2).
Case reports

FIG. 1. Basal ganglia calcification demonstrated by computerized axial tomography.

Discussion

Pseudohypoparathyroidism is confirmed in this patient by the classical biochemical findings and lack of renal response to exogenous parathyroid hormone. The radionuclide and histological evidence of increased bone turnover suggests skeletal responsiveness to parathyroid hormone, as is found in the variant of pseudohypoparathyroidism known as pseudohypparathyroidism described by Frame et al. (1972).

McKinney (1962) has described disorders of movement, including Parkinsonism, in both pseudohypoparathyroidism; clinicians should be aware that hypocalcemia can significantly exacerbate the features of Parkinsonism. Presentation in middle-age, absence of somatic features and absence of calcification of the basal ganglia on skull X-ray do not exclude a diagnosis of pseudohypoparathyroidism. Superiority of CAT in detection of basal ganglia calcification has been observed by Danziger et al. (1980), and increasing use of this technique will sometimes reveal unsuspected hypoparathyroidism, although other causes of calcification should be considered.

FIG. 2: Response of (a) plasma and (b) urine adenosine 3', 5'-cyclic monophosphate (cAMP) to i.v. bovine parathyroid hormone (200 u.). (a) x, patient; O, control. (b) •, patient; □ control.
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References


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