Idiopathic hypoparathyroidism with extrapyramidal and myopathic features

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SUMMARY A case of idiopathic hypoparathyroidism and basal ganglia calcification with extrapyramidal motor and myopathic features is described. The extrapyramidal motor and myopathic features disappeared when normocalcaemia was restored with treatment, suggesting that they resulted from hypocalcaemia.

Idiopathic hypoparathyroidism is an uncommon disease of childhood in which the typical neuromuscular disorders are tetany, fits, and mental retardation. Extrapyramidal and myopathic features have only rarely been described.

Case report
A 12-year-old boy presented with a 2-year history of...
sudden episodes of generalised tingling, followed immediately by uncontrollable muscle spasms with his limbs assuming bizarre postures, occasionally resulting in his falling to the ground. The episodes occurred 2 or 3 times a week and appeared to be more severe and more frequent after exercise.

He was the only child of healthy unrelated parents. His early growth and development had been normal. When aged 3, he was noted to have an abnormal gait attributed to coxa vara; there was some improvement with treatment, but he continued at times to walk stiffly on tiptoe. His parents felt that for 2 years he had lacked drive, but his school performance had remained satisfactory.

On examination, he was an intelligent boy of normal stature with no dysmorphic features. He had a mask-like facies, exhibited a poverty of movement, and walked with a slow stiff gait; he had strikingly increased muscle tone in all limbs without muscle wasting, tenderness, or weakness; deep tendon reflexes were diminished; he had difficulty in performing rapid repetitive movements, but he was not ataxic and had no sensory deficit. Chvostek's sign was positive and he exhibited chronic carpo-pedal spasm.

Investigations. Plasma calcium 1.37–1.83 mmol/l (5.48–7.32 mg/100 ml), phosphate 2.49–2.87 mmol/l (7.7–8.9 mg/100 ml), magnesium 0.66–0.70 mmol/l (1.6–1.7 mg/100 ml), albumin 44–49 g/l (4.4–4.9 g/100 ml), alkaline phosphatase 183–272 IU/l; urine calcium excretion 0.15 mmol/24 h (6 mg/24 h), phosphate excretion 10.21 mmol/24 h (0.32 g/24 h). Serum parathormone (PTH) 50 pg/ml (normal 250–680), serum 25 hydroxycholecalciferol 27.5 μg/ml (normal 5–30). Plasma creatine kinase (CPK) 13.0 μmol/ml per hour at 37°C (normal 0.25–3.6), alanine aminotransferase 23 IU/l, aspartate aminotransferase 22 IU/l. An Ellsworth Howard test showed a phosphaturic response to PTH. ECG showed a prolonged QTc of 0.49 seconds. An x-ray of skull was normal, but computerised axial tomography (CAT) showed dense calcification in both lentiform and caudate nuclei and in both cerebral hemispheres (Figure). EEG showed an excess of large amplitude slow activity without localising or paroxysmal features.

Blood count, ESR, blood urea and glucose, plasma electrolytes, creatinine, bilirubin, immunoglobulins, cortisol and ACTH, serum thyroxine and TSH, urine analysis, urine amino-acids, and faecal fat excretion were normal; no autoantibodies were detected.

A diagnosis of idiopathic hypoparathyroidism was made and treatment was started with dihydrotachysterol (DHT), with calcium and magnesium supplements, and aluminium hydroxide. There was a rapid response to treatment with complete relief of symptoms and signs when the plasma calcium rose above 1.9 mmol/l (7.6 mg/100 ml). Maintenance therapy with DHT 0.5 mg daily continues and the patient remains well.

Discussion

The patient had idiopathic hypoparathyroidism and basal ganglia calcification demonstrated by CAT. He exhibited extrapyramidal motor manifestations, in part related to exercise, which disappeared when normocalcaemia was restored. He also exhibited increased CPK activity to a level associated with myopathy which returned to normal when normocalcaemia was restored. While basal ganglia calcification will be seen more easily in the future by CAT, and therefore may be found more often in idiopathic hypoparathyroidism in childhood, extrapyramidal motor manifestations and raised CPK activity are both rare and do not appear to have been described previously in association with this condition.

Muenter and Whisnant (1968) reviewed the association between extrapyramidal motor manifestations and basal ganglia calcification, and found that the neurological deficit was usually reversible.
Half life of theophylline in the preterm baby with apnoeic attacks

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SUMMARY  Plasma theophylline levels were measured by an enzyme immunoassay method in 6 preterm babies. The method gave accurate and rapid results. The study showed a considerably prolonged half life of an oral preparation.

Xanthine derivatives have become established as a method of treatment of recurrent apnoeic attacks in the preterm baby (Lucey, 1975). However, because of the interpatient variation and prolonged half-life of theophylline in such babies (Giaocoia et al., 1976), it is essential that plasma levels be estimated frequently so that adequate doses of the drug can be maintained and serious side effects avoided.

We used an enzyme immunoassay method (EMIT, Syva) for determining plasma theophylline levels on at least as 10 μl plasma. It was known that plasma concentrations measured by either high pressure liquid chromatography or EMIT showed excellent agreement (Chang and Bastiani, 1977).

Patients and methods

Six preterm babies were studied. Details of gestation periods and birthweights are given in the Table. After one attack of apnoea lasting at least 25 seconds, a single oral theophylline preparation was given by a nasogastric tube (theophylline BP as sodium glyconate salt 5 mg/kg). Plasma theophylline levels (μg/ml) were measured at 0, 2, 4, 8, 12, 24, and 48 hours. If a baby had a second attack of apnoea during the period he was withdrawn from the study.

Plasma theophylline levels were estimated by enzyme immunoassay using the EMIT-aad kit

Table  Details of the 6 preterm babies

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Arch Dis Child 1979 54: 236-238
doi: 10.1136/adc.54.3.236

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