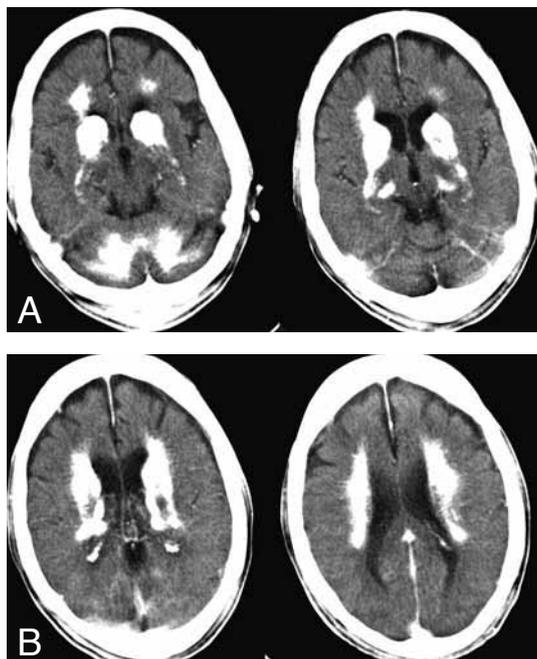


IMAGES IN CLINICAL RADIOLOGY



Idiopathic hypoparathyroidism as a cause of extensive intracranial calcification

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A 60-year-old male was referred to radiology department for head CT for evaluation of seizures of recent onset. CT showed extensive symmetric bilateral calcification in caudate nucleus, putamen, thalami, cerebellum and centrum semiovale (Fig. A,B). A radiological diagnosis of Fahr's disease was made. During hospital stay, patient developed muscular tetany. Serum calcium was found to be low (7.2 mg/dl; normal range 8.4-10.2 mg/dl). Further tests revealed low serum parathyroid hormone (PTH) level (3.60 pg/ml; normal range 15.0-68.0 pg/dl) and elevated phosphorus level. All possible causes of hypoparathyroidism were evaluated with no positive results. On the basis of clinical, hormonal and radiological findings a diagnosis of idiopathic hypoparathyroidism was made. Patient was started calcium and 1,25-dihydroxy vitamin D. Patient reported no episode of seizures during next two days of hospital stay. He was last seen at 3 months follow up with significant improvement of clinical symptoms and serum calcium level returning to normal range.

Comment

Fahr's is a rare neurodegenerative disease characterized by bilaterally symmetrical, extensive intracranial calcifications, especially in the cerebellum and basal ganglia. It is usually idiopathic, but can be associated with endocrine disorders such as hypoparathyroidism, pseudo-hypoparathyroidism, hypothyroidism and hyperparathyroidism (1). Other conditions associated with similar intracranial calcification pattern include infections, toxins (carbon monoxide, lead), metabolic syndrome (Cockayne's, mitochondrial disorders) and post chemotherapy.

Hypoparathyroidism is diagnosed with decreased serum parathyroid hormone and calcium level, increased serum phosphate. Common causes include renal insufficiency, chronic diarrhoea and alkalosis. Idiopathic hypoparathyroidism is a diagnosis of exclusion in a patient with low serum calcium and PTH level; excluding these and other causes of hypoparathyroidism such as surgery and autoimmune diseases (2).

Clinical signs and symptoms of hypoparathyroidism are due to hypocalcaemia and increased neuromuscular excitability resulting in tetany, paresthesia, seizures and calcium deposition leading to cataract and intracranial calcification. Intracranial calcification is more common and is seen early in idiopathic hypoparathyroidism. Latent tetany is diagnosed by eliciting Chovstek's and Trousseau's sign (2). Diagnosis is confirmed by biochemical analysis of serum calcium and PTH level. Treatment consists of oral or intravenous infusion of calcium and vitamin D.

Reference

1. Acou M., Vanslembrouck J., Deblaere K., Bauters W., Achten E.: Fahr disease. *JBR-BTR*, 2008, 91: 19.