

KNOWLEDGE APPLIED TO PRACTICE

APPLICATION DES CONNAISSANCES À LA PRATIQUE

DIAGNOSTIC CHALLENGE

Answer

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The correct diagnosis is hypoparathyroidism (a).

The CT scan of the head demonstrates diffuse intracranial calcifications, and on laboratory investigation the patient was found to be severely hypocalcemic (serum calcium was 1.33 mmol/L; normal range 2.10–2.60 mmol/L). His phosphorous was elevated (2.1 mmol/L; normal range 0.8–1.5 mmol/L) and, of course, his parathyroid hormone level (PTH) was low (0.4 pmol/L; normal range 1.5–7.6 pmol/L).

Intracranial calcification, especially in the basal ganglia, is a recognized complication of longstanding hypocalcemia due to hypoparathyroidism or pseudohypoparathyroidism.^{1–8} Diffuse calcification of the cerebral cortex, subcortical matter, basal ganglia, thalamus and cerebellum are

easily identifiable on the CT scan of this patient's brain (Fig. 1 and Fig. 2).

Metastatic carcinoma of the brain most commonly originates from a primary bronchogenic carcinoma. The appearance on CT scans of the brain is that of a low-density mass with surrounding ring enhancement at the junction of the grey and white matter.⁹

Tuberous sclerosis is a neurocutaneous disorder characterized by childhood epilepsy, intellectual impairment and dermatological manifestations. CT scans of the head typically reveal subependymal nodules or cortical "tubers" in the brain.¹⁰

The appearance of blood due to subarachnoid hemorrhage on CT scan is dependent upon the location of the aneurysm within the circle of Willis and the delay between



Fig. 1. Non-contrast enhanced CT scan of the head showing the dentate nucleus (A) and the vermis of the cerebellum (B).



Fig. 2. Non-contrast enhanced CT scan of the head showing the caudate (A), the globus pallidus (B), the putamen (C) and the thalamus (D), which are abnormally calcified in this image. Subcortical white matter is also diffusely calcified. The pineal gland (E) is normally calcified, as demonstrated here.

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the onset of symptoms and imaging. In general, the scan reveals increased density in the subarachnoid spaces, but it may also demonstrate an intraventricular or intraparenchymal hemorrhage, as well as hydrocephalus.¹¹

Commentary

Hypoparathyroidism is defined by low serum calcium and high serum phosphate with parathyroid hormone levels ranging from the undetectable to low-normal range. This is distinct from ineffective PTH action (pseudohypoparathyroidism), in which even mild hypocalcemia is associated with elevated PTH levels. Hence, in the setting of low serum calcium, failure to detect elevated PTH levels establishes the diagnosis of hypoparathyroidism.² Idiopathic hypoparathyroidism is diagnosed by the exclusion of known causes of hypoparathyroidism (neck surgery, autoimmune diseases, congenital and familial forms of this condition).

The signs and symptoms of hypoparathyroidism are related to hypocalcemia and increased neuromuscular excitability, which manifests as tetany, paresthesias and seizures.^{5,12} Bilateral symmetrical calcification of the basal ganglia, dentate nuclei and corona radiata is characteristic of chronic hypocalcemia secondary to hypoparathyroidism.^{3,4} The pathogenesis of intracranial calcification remains unclear.^{4,5}

Brain calcifications, even where extensive, do not necessarily lead to symptoms.¹ There are, however, case reports of extrapyramidal disorders such as parkinsonism, hemiballismus and choreoathetoid movements in adults with basal ganglia calcification.⁵⁻⁷ These conditions are often but not always reversible with correction of the hypocalcemia.⁶

Similarly, seizures in hypoparathyroidism are usually the sequelae of hypocalcemia.⁵ Treatment consists of supplementation with calcium and 1,25-dihydroxy vitamin D to prevent seizures and progression to extrapyramidal disorders.⁴ Hypercalciuria and nephrolithiasis are expected complications of long-term therapy with calcium and vitamin D analogues.⁴ Treatment with calcium has not been shown to prevent progression of the cerebral calcifications.

Conclusion

Chronic hypocalcemia was the cause of the patient's long-standing, intermittent paresthesia of the arms.⁸ Chvostek's sign (produced by tapping the skin over the facial nerve that is anterior to the external auditory meatus to elicit ipsi-

lateral contraction of the facial muscles) and Trousseau's sign (thumb adduction, metacarpophalangeal joint flexion and interphalangeal joint extension upon inflating a sphygmomanometer to 20 mm Hg above the systolic blood pressure for 3–5 min) were both present.^{2,13}

Treatment was initiated with calcium (intravenous and oral) and 1,25-dihydroxy vitamin D (Rocaltrol). The patient was admitted to hospital for 3 days and he suffered no further seizures.

For the Challenge, see page 367.

Competing interests: None declared.

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