Abstract

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Sepsis and Fahr's Syndrome

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Female, 67 years old, with hyperpyrexia

Past medical history: Fahr's disease with mild and constant progressive loss of autonomy in normal daily activities; previous removal of the pituitary gland for benign tumor with secondary hyponatremia.

Physical examination: Patient in comatose state, low back pressure ulcers. Dysphagia with nasogastric tube and PICC.

Clinical tests at admission: Severe anaemia (Hb 7.5 g / dl), hyponatremia (Na 119 mmol / l), positive occult blood test. ECG: sinus rhythm. CT of the skull: moderate signs of chronic vascular distress with diffuse hypodensity of the white matter of the radiated crowns and semi-oval centers. Bilateral calcifications of the nuclei of the base. EGDS: hypotonic oesophagus with incontinent cardia, without endoscopic signs of esophagitis. Hypotonic stomach with presence of decubitus ingests in correspondence of the body-fundus junction with apparently intact mucosa. Duodenal bulb covered with regular mucosa in the absence of signs of active or previous bleeding. Blood culture: E. Coli and S. aureus.

Removed PICC, practiced blood transfusion and specific antibiotic therapy. With the improvement of clinical conditions, PEG positioned therapy with Tolvaptan and normalization of hyponatremia.

Diagnosis: Generalized sepsis. Chronic cerebral vasculopathy in a patient with Fahr's syndrome. Bed rest with sacral pressure ulcers. SIADH anamnestic for hypophysectomy.

Fahr's disease is an idiopathic or familial neurodegenerative form characterized by symmetrical calcifications of brain structures, particularly basal ganglia and dentate nuclei of the cerebellum, with associated neuropsychiatric symptoms. Calcifications of the basal ganglia can be incidental findings (CT or MRI) in up to 20% of asymptomatic patients. The idiopathic forms are not associated with metabolic causes or other triggers. The secondary forms (Fahr's syndrome) are associated with several conditions: endocrine abnormalities of PTH (eg parathyroidism), other genetically determined conditions, and encephalitis and toxic substances. The physiopathology of S. of Fahr would be attributable to abnormalities of calcium-phosphorus homeostasis and alterations of the blood-brain barrier. It is caused by the association of a series of conditions: endocrinopathies, mitochondrial myopathies, dermatological and infectious diseases. Heterogeneous clinical manifestations: extrapyramidal symptoms and signs, memory and concentration alterations, movement alterations (Parkinsonism, chorea and tremor). Diagnostic criteria: evidence of bilateral calcification of the basal ganglia and of progressive neurological changes. Therapy: relief of symptoms and elimination of triggers.

Discussion: This case report has shown that patients with silent neurodegenerative conditions such as Fahr's disease, in the context of severe sepsis, may have an acute and rapidly progressive onset of the neurological symptoms themselves.