Letters to the Editor

Pseudotumoral neuro-behçet in a patient treated with anti-tumor necrosis factor alpha

Síndrome neuroológico seudotumoral de la enfermedad de Behçet en un paciente tratado con antagonistas del factor de necrosis tumoral

Dear Editor,

One of the main causes of morbimortality is the central nervous system condition caused by Behçet’s disease. It is classified in 2 main categories: nonparenchymal, causing vascular deterioration, and parenchymal, which develops like an aseptic meningoencephalitis. In some cases, there might be extensive brain lesions known as pseudotumoral neuro-Behçet’s disease. In 2012, Noel et al. published a compilation of 23 cases reported in medical literature, and they estimated that this presentation had a 1.8% prevalence among neurological manifestations. The most common histopathological finding was inflammatory perivascularitis. The typical image seen on magnetic resonance imaging (MRI) was a space-occupying lesion with mass effect, T1-hypointense enhanced by contrast, and T2-hyperintense; lesions were mostly located at the capsulolenticular, thalamus, brain stem, basal ganglia and cerebral cortex areas. We present the case of a patient who developed a severe, treatment-refractory, pseudotumoral syndrome.

Clinical case: 23-year old male diagnosed with Behçet’s disease in 2008, initially treated with prednisone 20–30 mg/day and colchicine, with partial response. In February 2011, he was given a treatment based on infliximab (IFX); 4 months later the patient had an adverse reaction, and the drug was changed for adalimumab (ADA). In October 2011, the patient started experiencing gait alterations, dizziness and vomiting, and had to be admitted. The MRI showed a pseudotumoral lesion at the right hemispheric, and less extensive injuries in the white substance at the right-sided inner capsule anterior arm, and in the area near mesial arm temporal structures (Fig. 1). In addition to ADA, the patient was administered oral azathioprine (1 mg/kg/day) and methylprednisolone (MP) by intravenous pulses (IV) (500 mg/day/5 days), with disease remission. A month later the patient was readmitted with fever, behavioural disturbances, hallucinations and short-term memory loss. The cerebrospinal fluid analysis evidenced aseptic meningitis without monoclonal expansion. The patient received new pulses of MP (500 mg/day/5 days), showing a good response.

Fig. 1. Magnetic resonance axial brain image: there is an area at the supratentorial compartment where signal disturbance pertaining the white substance adjacent to the left-side inferior horn is observed. There is another area in the posterior cavity with signal disturbance showing mass effect affecting right-sided cerebellar hemisphere. These lesions are T2-hyperintense with some internal heterogeneous areas.

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he was started on a treatment with tocilizumab (TCZ), and when corticosteroids were orally administered, symptoms reappeared. The brain MRI showed an inflammatory extensive lesion at the limbic system structures, more accentuated on the left side. A new MP cycle (500 mg/day/5 days) including IV cyclophosphamide IV (500 mg/month) was administered. After a month, the patient developed right-sided hemiparesis and suffered deterioration of his supratentorial and infratentorial lesions, as shown by the MRI. A new MP cycle (1 g/day/5 days) including rituximab was administered, and the patient received 5 sessions of plasmapheresis. Due to the negative response to the treatment, a brain biopsy was performed, causing a 5 cm × 3 cm intraparenchymal bruise above left-sided basal ganglia. The histological study showed reagent-like changes; there were no signs of encephalitis or atypical lymphoid elements. Two days after completing the brain biopsy, the patient had an epileptic seizure with a prolonged post–critical period, having to be admitted to the intensive care unit. After 5 months of undergoing an immunosuppressant treatment, the patient passed away due to an intraparenchymal haemorrhage.

Discussion: no medical strategy to treat neuro-Bechter’s disease has been ratified in controlled clinic trials to date. Most patients with pseudotumoral lesions received a treatment with high doses of steroids along with immunosuppressants, and in 2011, the case of a patient who obtained clinical remission after being treated with biological therapies (daclizumab, IFX and TCZ) was reported. The neurological symptoms experienced by our patient had first appeared during the treatment with tumour necrosis factor (TNF) blockers, a drug that has been associated with the development of demyelinating lesions that, typically, improve when the treatment is suspended. The brain lesions sustained by our patient were extensive and persisted after suspending the anti-TNF treatment; the corticosteroids and immunosuppressants administered proved to be inefficient. In the series compiling 23 cases, 2 patients passed away (8.7%) and 7 (30.5%) had some sort of sequelae, confirming that pseudotumoral neuro-Bechet is one of the presentations of the Bechet’s disease, increasing the risk of suffering severe sequelae and threatening life.

References

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Intrapericardial paraganglioma associated with mutation in succinate dehydrogenase enzyme gene

Paraganglioma intrapericárdico asociado a la presencia de mutación en el gen de la enzima succinato deshidrogenasa

Dear Editor,

Paragangliomas are neuroendocrine tumours derived from the chromaffin tissue of the autonomic nervous system lymph nodes. Most frequently located in the abdominal area, and less frequently in the thorax, pelvis or the cervical area. They may be functional secreting catecholamines, giving rise to adrenergic symptoms that might be potentially serious.

We present the case of a 34-year old female referred by her primary care physician with a hypertensive crisis coinciding with headache. The patient did not have a family history of arterial hypertension (AHT) or any other relevant medical condition. The only chronic drug treatment she received was lorazepam for insomnia. During the previous year, she had multiple visits to the Emergency Department with sudden onset of headache, sweating, heart racing and agitation, lasting from one minute to an hour, along with occasional chest pain and nausea. These symptoms had been diagnosed as anxiety, migraine and acute pericarditis.

Over the last month, these episodes became more frequent and intense, presenting blood pressure (BP) up to 200–100 mmHg, together with a heart rate (HR) of 110 bpm. The general laboratory test showed mild iron-deficiency anaemia, but her other parameters, including calcium, were normal. The specific laboratory test revealed highly elevated plasma levels of norepinephrine (INN) and normetanephrine (NMN), and slightly elevated levels of dopamine (DA) [adrenaline 31.3 pg/ml [NR <80], norepinephrine >2700 pg/ml [NR <446], dopamine 290.8 pg/ml [NR <150], metanephrine 15 pg/ml [NR <90], NMN 972 pg/ml [NR <180]] and urine of 24 h [adrenaline 6.63 μg [NR <27], INN 1.279 μg [NR <97], DA 1535 μg [NR <500], metanephrine 83 μg [NR <320], NMN 2633 μg [NR <390)]. An abdominal and pelvic CT angiography was ordered, showing no changes, with normal parameters for both adrenal glands, and, to complete the localization study, a cervical-thoracic CT scan was made, revealing an heterogeneous and very vascularized 5 cm mass located in the middle mediastinum, with epicentre at the subcarinal area. The functional study with ¹¹C-MIBG showed a unique mediastinal capture, confirming the paraganglioma diagnosis. An alpha-blocker treatment with phenoxybenzamine 10 mg/day was started, progressively increasing the dose up to 60 mg/day, subject to BP figures, and 7 days later, it was coupled with a beta-blocker treatment with propranolol, which was later replaced with atenolol, up to 50 mg/12h to control HR. Moreover, it was recommended that the patient should drink liquid and consume salt. A month later, the patient underwent a surgery whereby the intrapericardial mediastinal mass was extirpated by means of a right–side posterolateral thoracotomy, without reported incidents during or after the surgery. The pathological anatomy confirmed the presence.