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Small-fibre polyneuropathy in Whipple's disease

Afección neurológica en la enfermedad de Whipple en forma de polineuropatía de fibra fina

Dear Editor:

Whipple's disease is a rare multisystemic disease caused by the bacterium *Tropheryma whippelii*. It typically causes rheumatic and digestive affectation, along with frequent neurological symptoms in the final stage of the disease[1,2], especially in the form of supranuclear ophthalmoplegia, cognitive impairment and confusion. In the neurological clinic, the affectation of the peripheral nervous system is one of the least described. We present a patient with fine fibre polyneuropathy caused by Whipple's disease.

The patient is a 58-year-old man, with no toxic habits, with a history of chronic seronegative oligoarthritis of more than six years of evolution, who attended the neurology service due to plantar dysesthesia with 1 year of evolution and a progressive course. On examination, segmental muscular balance was normal in both lower and upper extremities and bone-tendon reflexes were within normality. In terms of sensitivity, we detected sock hypalgesia, along with distally decreased pallesthesia.

Analysis revealed that the blood count, renal and liver function, clotting, tumour markers, thyroid function, autoantibodies (ANA, ENA ANCA, ECA, anti-smooth muscle), serology for hepatitis, syphilis, HIV, protein electrophoresis, cryoglobulins, vitamin B12 and folic acid were within the normal results. Only an erythrocyte sedimentation rate (ESR) of 70 mm in the first hour stood out, maintaining similar values in the following analytical controls.

Neurography showed a slight decrease in amplitude of sural sensory potentials (left, 10.73 μV; right, 8.69 μV. Reference values, according to Kimura, 17.2 ± 6.7 μV), with the rest of the study within the normal range, including coaxial electromyogram.

Given that they would explain the clinical manifestation of xerophthalmia, a Shirmer test and salivary gland scintillography were conducted, with negative results. A biopsy of subcutaneous fat was also performed, with no detectable amyloid deposits.

The clinical picture followed a sluggish course, with increasing dysesthesia and poor symptom control despite combination therapy (capsaicin, phenytoin, valproic acid, gabapentin, baclofen). A new, control electrophysiological study was performed again after one year, with no significant changes.

Eighteen months later, the patient consulted for continuous abdominal pain with vomiting and diarrhoea, having lost 10 kg. We performed a small intestine biopsy by endoscopy and detected macrophage infiltration with PAS-positive inclusions. We performed a sural nerve biopsy, which showed a reduction of unmyelinated fibres with preservation of the medium- and large-calibre fibres in the absence of inflammatory and vascular changes (anatomopathological method: fixation in glutaraldehyde, after fixation in osmium tetroxide, dehydration in ethanol and propylenoxide, inclusion in araldite).

Under the diagnostic guidance of Whipple's disease affecting joints, digestive tract and peripheral nervous system in the form of fine fibre polyneuropathy, we initiated treatment with intravenous ceftriaxone for 15 days and cotrimoxazole for 1 year. As for the analysis, ESR normalised and there was a marked improvement of digestive symptoms, although arthralgias and dysaesthesia persisted.

The literature reflects about 200 cases of Whipple's disease with neurological affectation; of these, we have found in approximately 5% peripheral neuropathy described in different forms: sensorimotor mixed polyneuropathy[3,4,5] (the degree of association in one of them is arguable because the patient was also a severe alcoholic[4]), polyneuropathy (predominantly motor)[5], bilateral wrist-drop[6], CPE paralysis[1,4] (the latter probably related to severe weight loss). Three of these patients underwent sural nerve biopsy: one showed a non-specific perivascular inflammatory infiltrate[6]; the second, a moderate loss of myelinated and unmyelinated fibres, with some macrophages, but without any bacilliferous inclusions[3], and the third showed bacilliferous particles, but no PAS-positive inclusions[7].

After reviewing the literature, ours seems to be the first reported case of fine-fibre polyneuropathy confirmed by sural nerve biopsy. Despite the absence of PAS-positive inclusions in our biopsy as well, it could not be attributable to other causes because of the negativity of the rest of the study. Also pointing to the prior manifestation of this disease is the fact that ESR was persistently elevated and became normalised after specific antibiotic treatment.

In the first neuropathies described in Whipple's disease, it was considered that they were secondary to hypo-absorption[4]. However, there were subsequent cases

described in which the neuropathy preceded the abdominal clinical manifestations by years. In our patient, the clinical picture of polyneuropathy began two years before the abdominal manifestations and no nutritional deficiency was indicated. Various forms of peripheral neuropathy have been reported in other chronic inflammatory intestinal diseases, with an unclear relation to hypo-absorption.

Presentations

This clinical case was presented at the meeting of the Catalan Society of Neurology in 2007.

Financing

This work has not received any kind of financing.

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Klüver-Bucy syndrome secondary to medulloblastoma metastasis

Síndrome de Klüver-Bucy secundario a metástasis de meduloblastoma

Dear Editor:

We describe the case of a 33-year-old male with a history of medulloblastoma diagnosed at age 29, treated at the time with surgical resection, cranial and neuraxis radiation therapy and chemotherapy. Four years later, he began to manifest a hypersexual behaviour, hyperorality, disinhibition and placidity; he was taken to the emergency department for impaired alertness. On admission, he was sleepy, with abulia and psychomotor agitation, uninhibited, with delusional and manic ideas and bilateral papilledema; the remainder without alterations. The imaging study showed two symmetrical lesions in the anterior temporal pole, with uptake of contrast medium. They were removed in two surgical procedures; the histopathological study led to the diagnosis of temporal metastases of medulloblastoma (fig. 1). He suffered a remission of neuropsychiatric symptoms.

Heinrich Klüver and Paul Bucy described behavioural changes that could be generated in monkeys by performing bilateral temporal lobectomy. The first case of Klüver-Bucy syndrome (KBS) in humans was published in 1947[1,2]. The most important symptoms of human KBS are visual agnosia, placidity, emotional numbing, increased sexual activity, hyper-metamorphopsia, increased oral behaviour and bulimia.

Numerous causes of KBS have been described. The most frequently associated causes are traumatic injuries, epilepsy and infections of the central nervous system, especially with temporal lobe involvement[3], although there are cases of degenerative, vascular and metabolic lesions. A rare manifestation of tumour lesions, there are only two cases documented in the literature, one with bilateral temporal oligodendroglioma[4] and another with bitemporal arachnoid cyst[5]. To our knowledge, there is no other case of KBS caused by bitemporal metastases of medulloblastoma in the literature.

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