CASE REPORT

D evic disease: a case report

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Abstract

Objective: to report a case of Devic disease, emphasizing its diagnosis, in addition to reviewing the medical literature.

Description: male, six-year-old patient suddenly developed weakness in lower limbs, with resolution during hospital stay. However, as the weakness disappeared, loss of vision occurred. The symptoms were reverted after the use of prednisone.

Comments: the diagnostic and therapeutic approach was similar to that used in other cases reported by different reference centers. In other words, clinical diagnosis and prednisone therapy were used, with the complete improvement of symptoms. However, there is still some controversy surrounding its etiology and relationship with other demyelinating diseases, such as multiple sclerosis.


Introduction

Devic disease or neuromyelitis optica is a rare disorder of unknown etiology that belongs to the category of demyelinating diseases of the central nervous system.1 In most cases, a specific etiology is not identified; however, an immunological mechanism of tissue damage seems probable.2 Studies show the presence of anti-Epstein-Barr virus antibody in patients affected by the disease3 and a possible association with TBC.4 Devic disease usually occurs in children. Some authors relate it to a form of multiple sclerosis, and others, to acute disseminated encephalomyelitis.1 Demyelination is secondary and may be related to multiple factors, such as antirabies vaccine or intoxication, among others.1 Herein, we describe a case of Devic disease, presenting all the diagnostic tools, therapeutic conduct, and follow-up of the patient during one year after diagnosis.

Case report

In March 99, a white, six-year-old child was admitted to the emergency room of the Hospital de Clínicas de Porto Alegre (HCPA) with suspicion of Guillain-Barré syndrome.
Two days before, the patient had presented with myalgia, diffuse headache, and walking difficulty, which progressed into walking disability, even if aided; two episodes of urinary retention were also reported. Other intercurrent diseases were not observed, except for upper airway infection in the previous week. The patient had a previous history of nocturnal enuresis. The physical examination revealed weakness of the lower limbs (without spontaneous mobility), lower limb areflexia, reduced vibratory sensitivity in right lower limb, and pain when the lower limbs were palpated. Funduscopic examination was normal. The results of lab exams were: normal urinalysis, CSF with slight increase in the amount of proteins (65 mg/dl) and hemogram with leukocytosis (12,500 rods, 64% segmented, 9% eosinophils, 7% monocytes and 24% lymphocytes). At first, there was suspicion of Guillain-Barré syndrome with not so characteristic CSF.

While in hospital, the patient presented with urinary retention once again, and required a urinary catheter. A new examination of the ocular fundus revealed blurring in the lower papillary region in both eyes. Cranial CT scan revealed right-sided hypodensity in the frontal and temporal region. The CT scan of the spinal cord was normal. On the third day in hospital, improvement of lower limb weakness was observed, level II was reached, and deep reflexes reappeared. New CSF with normal values: nonreagent toxoplasmosis IgM, reagent IgG. Nonreagent LUES IgG, negative bacteriological exam, ADA 2.8 U/L, and absence of fungi. Normal hemogram, gas blood analysis and electrolyte results were obtained. Normal sensitive and motor electroneuromyography, incompatible with Guillain-Barré syndrome.

On the fifth day in hospital, patient’s eyesight was affected. Neurological exam: strength level IV in lower limbs and level V in upper limbs, presence of phasic, myocytic, and symmetric reflexes in upper and lower limbs. Bilateral flexor cutaneous-plantar reflex. Unaltered sensitivity. On ophthalmologic exam, slow pupillary light reflex of the right eye, and wandering look were observed. Examination of the ocular fundus: papillary edema on the left side, and retinal hemorrhage suggestive of neuritis. Clinical examination compatible with optic encephalomyelitis, for which a loading dose of prednisone 2 mg/kg was administered, improving the symptoms on the following day. EEG: alterations associated with diffuse cortical involvement (moderate), slow focal alterations compatible with focal involvement of the fronto-temporal areas of the left hemisphere. At the end of the tenth day, the patient was discharged from hospital, with some spasticity and diagnosis of optic neuritis, encephalitis, and myelitis (Devic disease). Optical fundus with persistent hemorrhagic alterations and papillary blurring. Therapeutic conduct: use of prednisone 2 mg/kg/day for two weeks; after that, the dose should be reduced to 1.5 mg/kg/day, twice a day; and to 1 mg/kg/day every morning, thereafter. Outpatient follow-up: use of corticosteroids for five months until neurological exam yielded normal results. Ophthalmologic routine exams did not show any abnormalities.

Discussion

Devic disease is first characterized by impairment of visual acuity and spinal cord function. Both disorders are often present some days after the onset of the disease, and are usually combined for two weeks. Optic neuritis may occur as papillitis or retrobulbar neuritis. More often than not, it initially occurs after myelitis. The most common medullary syndrome is motion-sensitive crural paraplegia, with a well-established level of sensitivity. At the beginning, it is flaccid, becoming spastic over time. Sphincter disorders are constant and occur at the early stage. Visual acuity of both eyes is greatly impaired, but may develop unilaterally. Central scotoma is normally present. Edema is likely to occur, and the papilla may look normal or enlarged. Impairment of visual acuity progresses within days or in a few weeks. Ophthalmoplegia is rare, but Horner syndrome may be present. Symptoms may totally improve, and visual acuity and spinal cord function may be partially reestablished. Death may occasionally occur at acute stages. The anatomical and pathological exam shows injured spinal cord, and optical nerves, but usually uninjured encephalon. Myelin loss and slight axon degeneration are observed. Massive edema, with foci of perivascular demyelination, may be present; sometimes, confluence of medullary lesions may occur, which characterizes necrotizing transverse myelitis.

The diagnosis is essentially based on clinical symptoms. It is necessary that transverse myelitis with well-established level of sensitivity and optic syndrome, papillitis (intrabulbar neuritis) or retrobulbar neuritis be present. CSF may reveal high levels of gamma-globulin. Compressive medullary lesions should be ruled out by imaging exams. Nuclear magnetic resonance shows the presence of lesions in the white matter, similar to those occurring in multiple sclerosis, in addition to the saltatory progression of symptoms. Thus, one may conclude that Devic disease is a variant of multiple sclerosis instead of a unique disease, although otherwise suggested by some studies.

The use of prednisone has been recommended as treatment and, in some studies, the total remission of symptoms was obtained. Our patient received this treatment and presented excellent results. Mandler et al. (1998) did not find recurrent symptoms for eighteen months in patients treated with prednisone and azathioprine during two months. Rilling et al. (1999) showed that the use of methylprednisolone improved clinical symptoms for a while, and cyclophosphamide was used to treat recurrent symptoms. The prognosis is worse when necrotizing myelitis is present; however, there has been excellent prognosis among pediatric patients. Retrospective studies showed that most patients developed severe disability and respiratory insufficiency during several decades.
follow-up of our patient (one and a half year after the episode) showed that the neurological exam remained normal. The clinical symptoms combined with the findings of the ocular fundus exam allowed us to diagnose optic neuromyelitis. The initial suspicion of Guillain-Barré syndrome was ruled out since no laboratory and neurophysiological evidence was obtained. Therefore, we conclude that the possibility of optic neuromyelitis should always be considered whenever clinical symptoms such as those observed in our patient are present. Nevertheless, the underlying mechanisms of Devic disease need to be further investigated and better understood so that we can associate it or not with multiple sclerosis, and standardize its treatment. This is only possible if patients are followed up for several years.

References

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