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West syndrome and multiple sclerosis association. About a case

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ABSTRACT

Introduction. West syndrome is a rare and severe infantile epileptic encephalopathy, beginning around the age of six months, characterized by a classic electro-clinical triad. This is a pathology totally different from multiple sclerosis (MS) which is a demyelinating disease of the central nervous system caused, affecting young adults, especially females. The association of these two pathologies has never been described.

Observation. We report here an exceptional presentation of MS in a 14-year-old girl with a history of West syndrome. She had normal development until the age of six months, when she began to have flexion spasms. The diagnosis of West syndrome was made with a normal MRI. The infantile spasms disappeared after treatment with vigabatrin and adrenocorticotropic hormone (ACTH). It had generally progressed to Lennox Gastaut encephalopathy, with delayed psychomotor development and epileptic sequelae. At 14, she presented with left hemiparesis within a few days. A cerebral MRI showed multiple nodular hyperintensities of the supra and infratentorial white matter, with the presence of an active lesion, fulfilling the diagnostic criteria for multiple sclerosis. CSF analysis was normal. Anti-AQP4, anti-MOG, anti-NMDA and anti-GABA (AB) antibodies were absent in the blood. Antibodies against HIV and viral hepatitis were. Biotinidase activity and autoimmunity tests were correct. The patient received high doses of methylprednisolone IV (1g/day) for three days with remarkable clinical improvement after 15 days.

Discussion. MS is a complex and heterogeneous central nervous system (CNS) demyelinating disease. It is not uncommon for epilepsy to be the first symptom of multiple sclerosis. Seizures, on the other hand, are more common after disease progression. Although the disease is characterized by inflammatory lesions of the white matter, various neuropathological and radiological studies have shown that the disease also affects the grey matter. Several studies have shown that seizures are three to six times more common in MS patients than in the general population. Even though MS can start with epilepsy and a seizure may be the only symptom of a relapse of MS, it is still not known whether the two diseases coexist or whether MS predisposes to seizures.

Conclusion. The association of these two totally different pathologies can lead us to say that the mechanism of multiple sclerosis may begin in childhood and that the clinical signs appear in adulthood.

Keywords
West syndrome, multiple sclerosis, inflammation

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INTRODUCTION

West syndrome is a rare and severe age-related epileptic encephalopathy of infancy comprising a triad of infantile spasms, hypsarrhythmia and psychomotor delay. 50% to 90% of patient evolve

to other syndromes mostly Lennox Gastaut syndrome, whereas multiple sclerosis (MS) is a central nervous system demyelinating disease caused by an autoimmune process. It is highly prevalence among young female adults. Their presence with a single patient has never been described.

We herein report an exceptional presentation of MS in a 14-year-old girl child with a childhood history of West syndrome.

Observation

A 14-year-old_female child presented with rapidly onset left hemiparesis. Concerning her past medical history, she was born into a second-degree consanguineous marriage. She had a history of intrauterine growth restriction (IUGR) and low birthweight in a normal delivery. Nevertheless, she had normal development until the age of six months, when she started having flexion and extension spasms occurring in clusters, usually shortly after waking up.

The diagnosis of West syndrome was established. The infantile spasms disappeared after three years of treatment with vigabatrin and adrenocorticotropic hormone (ACTH). It had typically progressed to Lennox Gastaut encephalopathy, with psychomotor developmental delay and epileptic sequelae.

At the age of four, she presented stereotyped behaviours, most commonly involving hands. Therefore, the diagnosis of Rett syndrome has been suspected, and it was ruled out by a negative genetic study. A brain MRI at this age was without abnormalities.

Her epilepsy was relatively stable under lamotrigine and sodium valproate with motor and cognitive sequelae. Interestingly, she was able to walk at the age of 8 years after intensive motor rehabilitation and was relatively autonomous in day-to-day tasks.

At the age of 14, she presented a left hemiparesis worsening over few days. On examination, we found a conscious, very agitated and irritable child with incessant cries and tears, retrognathism, microcrania, low hair implantation, stereotyped hand rubbing movements, generalized hypotonia and left hemiparesis. A brain MRI showed multiple nodular and bilateral T2/FLAIR hyper signals and T1 hyposignals in the supra and subtentorial white matter, with the presence of one active lesion in the left oval centre (Figure 1).

CSF analysis with iso-electro-focalisation showed a proteinorachia at 0.33mg/l with the absence of the oligoclonal band (OCB). The anti AQP4, anti-MOG, anti NMDA, and anti-GABA antibodies (AB) were absent in the blood. Antibodies to HIV and viral hepatitis were negative. The biotinidase activity and autoimmunity tests were correct.

The patient received high doses of IV methylprednisolone (1g/day) for three days with a remarkable clinical improvement after 15 days.

Five months later, her motor symptoms had worsened with the recurrence of her left hemiparesis. A brain and spinal cord MRI showed a significant increase in lesion load with the presence of at least eight active lesions as well as multiple spinal cord lesions (figure 2). Consequently, treatment was initiated, including an IV infusion of methylprednisolone (1 g/d for five days), followed by an IV Ig course (25mg/d for three days) with a favourable response. Another IV Ig course was given after one month, and the girl was remarkably stable.

Six months after that, the patient had another relapse, which was made of left arm weakness with an exacerbation of her seizures. A brain MRI was performed, showing the presence of multiple active lesions (figure 3). She was then admitted for an IV infusion of methylprednisolone (1g/d for three days) followed by monthly repeated courses of IV Ig as the diagnosis of MOGopathy was highly suspected.

Under this treatment, the patient's evolution was favourable, and he has been free of relapses ever since.

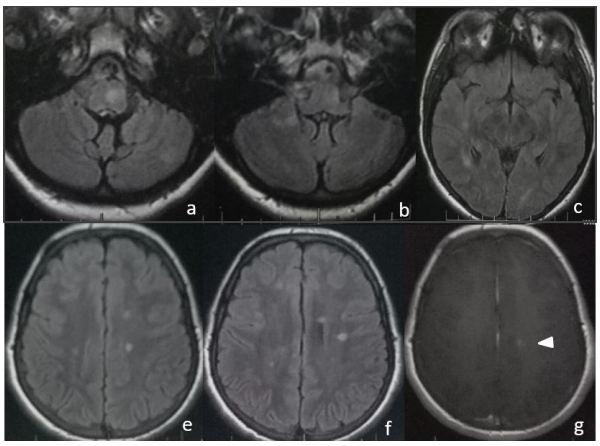

Figure 1. Brain MRI showing multiple FLAIR hypersignals in sub and supra tentorial white matter.

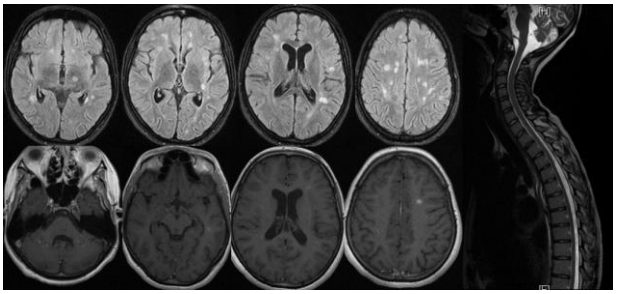

Figure 2. Brain and spinal cord MRI after 2 months, showing an increase in the number and volume of the lesions.

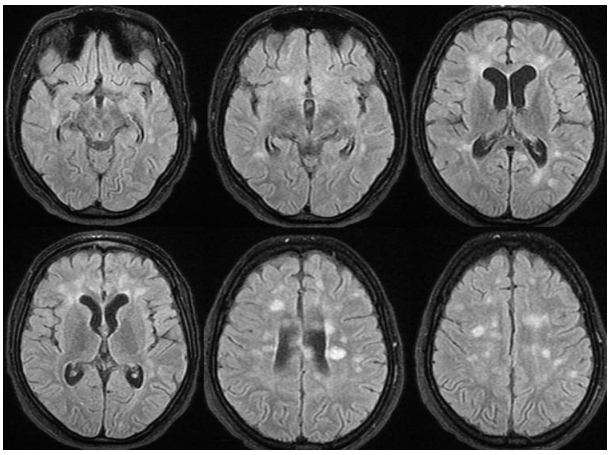

Figure 3. Brain MRI after 11 months from the first relapse.

Discussion

Multiple sclerosis (MS) is a central nervous system (CNS) demyelinating disease that is both complex and heterogeneous.

It is not rare for epilepsy to be the first symptom of multiple sclerosis. Seizures, on the other hand, are more common after the start of the condition (1). Although the disease is characterized by inflammatory lesions in the white matter, various neuropathological and radiological studies have shown that the disease also affects the grey matter. Several studies have showed that seizures are three to six times more common among MS patients than in the general population (2) (3). Even though MS can start with epilepsy (4) and that a seizure may be the only symptom of an MS relapse (5), it's still unclear whether the two diseases coexist or whether MS predisposes to seizures (6).

On the other hand, infantile spasms initially discovered by William James West in his own son in 1841.

The term Infantile spasms syndrome (ISs) now refers to an epileptic syndrome that affects children under the age of one year (rarely older than two years), with clinical spasms that usually occur in clusters, with hypsarrhythmia as the most common EEG finding. The spasms that are frequently associated with developmental arrest or regression. The name West syndrome (WS) refers to a subset of ISs marked by clustered spasms and hypsarrhythmia on an EEG, as well as delayed brain development or regression.

The pathophysiology of infantile spasms isn't completely understood. It is well-known that partial seizures can progress to spasms, as evidenced by invasive and surface EEG recordings, as well as the peculiar PET (positron emission tomography) finding of hypometabolism of deep grey structures and the brain stem in children with spasms, all support the cortical-subcortical interaction hypothesis (7). The therapeutic response to hormonal therapy, on the other hand, has shown a role for the hypothalamic-pituitary axis and immunological systems. The Developmental - desynchronization model better explains the narrow age of onset as well as the developmental effects (7).

Conclusion

The association of West syndrome and multiple sclerosis has never been described, and the common point is neuroinflammation, which can open perspectives on the understanding of the physiopathological mechanisms of these two pathologies, and can be an argument that the substance gray is not spared from multiple sclerosis.

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