IV and V cranial nerves dysfunction as initial manifestations of superficial siderosis of the central nervous system

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Superficial siderosis of the central nervous system is a rare disorder characterized by the deposition of hemosiderin in the subpial layer of the central nervous system. It is caused by chronic and insidious hemorrhages in the subarachnoid space, which are neurotoxic particularly to the cerebellum, brainstem and spinal cord. Clinically, the most frequent features are sensorineural hearing loss, cerebellar ataxia and myelopathy [1-3]. To our best knowledge trigeminal neuropathy has never been reported in superficial siderosis and there are only two case reports of superficial siderosis presenting with trochlear palsy [1].

We report the clinical case of a 79-year-old man who was admitted due to recurrent symptoms of nausea, vertigo and gait unsteadiness, which had worsened recently and had become incapacitating.

Besides long lasting dyslipidemia and hypertension, his past medical history was relevant for the onset in 2010 of episodes of left facial paresthesias affecting all three divisions of trigeminal nerve that alternated with dysesthesias in the same distribution. The diagnosis of atypical trigeminal neuropathy was made at the time. He was treated with gabapentin with partial improvement. Additionally, in 2011 the patient had recurrent episodes of vertical diplopia exacerbated by down-gazing. The ophthalmologist did the diagnosis of right trochlear nerve palsy with remission in a few weeks.

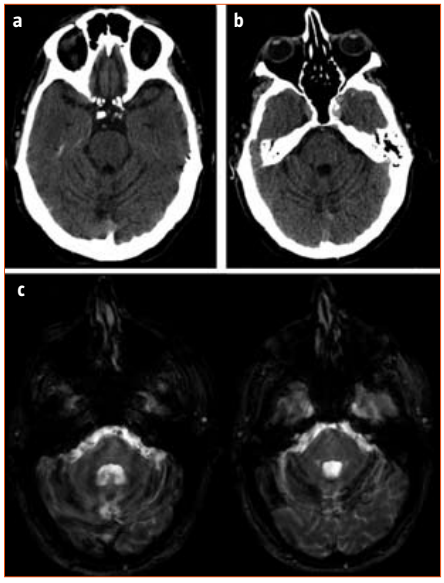

Figure. Head computerized tomography in 2010 (a) and 2014 (b), and brain magnetic resonance imaging in 2014, T_2* weighted images (c).

The current neurological examination disclosed neurosensorial bilateral hypoacusis, left superior limb and bilateral lower limb ataxia without significant worsening with eyes closing and cerebellar ataxic gait.

The brain magnetic resonance imaging revealed extensive hypointensities on FLAIR and T_2* throughout the surface of the hemispheric cerebellar folia bilaterally, vermis and some occipital gyri (Figure). There was also diffuse cerebellar atrophy. This cerebellar atrophy was already present in a brain computerized tomography done at the time of the atypical trigeminal neuropathy diagnosis in 2010 (Figure). A diagnosis of superficial siderosis of the central nervous system was therefore made. No aneurysm, arteriovenous malformation or dural fistulas were found in a cerebral digital subtraction angiography. No other causes for the trigeminal and trochlear involvement reported earlier were found in magnetic resonance imaging or in cerebral digital subtraction angiography, namely vascular nerve compressions.

Pathophysiologically, the tissue damage in superficial siderosis occurs as a result of an active glial process of conversion of heme to ferritin and hemosiderin that requires the presence of central myelin [4]. The susceptibility to cerebellar cortex lesion has been explained by the presence of a specific type of astrocytes, the Bergman glia, and by the abundance of microglia, which predispose to hemosiderin accumulation [4]. The first and second cranial nerves are frequently involved because they are part of the central nervous system. The eighth cranial nerve is also preferentially affected because the transition from central to peripheral myelin in this nerve occurs in the acoustic canal, 10-15 mm away from its emergence in the pons [4,5]. The remaining cranial nerves are not prone to siderotic deposition since the transition to Schwann cells is only 0.5-1 mm away from the brainstem or spinal cord [4]. Reviewing the literature, we found only two cases of trochlear nerve palsy and no cases with trigeminal nerve dysfunction. Although the absence of cases presenting with trigeminal neuropathy, there are two reports of asymptomatic prominent hemosiderin deposition in trigeminal nerves identified in magnetic resonance imaging [6,7].

The cerebellar atrophy was already present in a brain computerized tomography performed in 2010. Since no other cause for cerebellar atrophy was found, it is fair to deduce that the process of hemosiderin accumulation in the central nervous system had already begun at the time of the onset of atypical trigeminal neuropathy and trochlear nerve palsy.

This clinical case is highly suggestive that the previous IV and V nerves impairments were the initial manifestations of superficial siderosis, which constitutes a novel presentation of this disease.

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