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# Involvement of peripheral III<sup>rd</sup> nerve during MS: report of a new case and discussion of the underlying mechanism.

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## Abstract:

Multiple sclerosis (MS) is a chronic disorder that affects the central nervous system myelin. However, a few radiological cases have documented an involvement of

peripheral cranial nerves, within the subarachnoid space, in MS patients. We report the case of a 36-year-old female with a history of relapsing remitting (RR) MS who consulted for a subacute complete paralysis of the right III<sup>rd</sup> nerve. MRI examination showed enhancement and thickening of the cisternal right III<sup>rd</sup> nerve, in continuity with a linear, mesencephalic, acute demyelinating lesion. Radiological involvement of the cisternal part of III<sup>rd</sup> nerve has been reported only once in MS patients.

Radiological involvement of the cisternal part of V<sup>th</sup> nerve occurs more frequently, in almost 3% of MS patients. In both situations, the presence of a central demyelinating lesion, in continuity with the enhancement of the peripheral nerve, suggests that peripheral nerve damage is a secondary process, rather than a primary target of demyelination.

### **Case report:**

We report the case of a 36-year-old woman with an 18-year history of RR MS. She has been treated by interferon for the last 4 years, without any clinical relapse or MRI activity. The last relapse in 2010 left her with an Expanded Disability Status Scale (EDSS) of 2.0 due to sensory deficit of the lower limbs. In November 2015, she presented with a painless binocular vertical diplopia associated with right upper eyelid ptosis. Clinical examination revealed a complete palsy of the right III<sup>rd</sup> nerve (Figure 1A-D), with a vertical misalignment of the eyes in primary gaze, a limitation of elevation, adduction and depression of the right eye and a discrete enlargement of the right pupil. Visual acuity was normal for both eyes. There was no other new neurological deficit. Brain MRI showed a small, T2 Fluid Attenuated Inversion

Recovery (FLAIR) hyperintense, contrast-enhanced mesencephalic lesion compatible with acute demyelination (Figure 2A-C, arrowheads). This lesion was located at the inner side of the right cerebral peduncle, next to the root exit zone (REZ) of the right III<sup>rd</sup> nerve, which was enlarged and enhanced after contrast injection, from the REZ to the cavernous sinus (Figure 2A-C, arrows). MRI also demonstrated multiple demyelinating lesions of the corpus callosum and deep white matter (Fig 2D) without any post-gadolinium enhancement. The carotid arteries had a normal appearance (not shown). There was no compressive lesion on the right III<sup>rd</sup> nerve. The patient was treated by intravenous methylprednisolone (1g/day for 3 days) with a complete recovery of symptoms within 2 months of onset.

### **Discussion:**

MS is characterized by a demyelination of the central myelin, produced by oligodendrocytes. Thus, MS lesions should not affect peripheral cranial nerves, as they contain peripheral myelin produced by Schwann cells. Only one other case of radiological involvement of peripheral, cisternal III<sup>rd</sup> nerve in MS has been reported in the English-language literature (1). As in our case, there was a demyelinating lesion located in the cerebral peduncle, close to the emergence of the III<sup>rd</sup> nerve, with continuity between the enhancement of the demyelinating lesion and the enhancement of the cisternal III<sup>rd</sup> nerve. In both cases, the III<sup>rd</sup> nerve paresis was complete, with both intrinsic and extrinsic involvement. In our case however, pupillary involvement was relatively mild, which was not in favor of an extrinsic compression, rather suggesting an intrinsic damage of the III<sup>rd</sup> nerve. Cisternal III<sup>rd</sup> nerve enhancement

has been reported in other conditions, including infectious meningitis (tuberculosis, Lyme disease), carcinomatous meningitis, lymphoma and leukemia. Cisternal III<sup>rd</sup> nerve enhancement related to MS is exceptional, however, cisternal enhancement has been frequently observed in the V<sup>th</sup> nerve in MS patients (2,8-2,9% of cases) (2,3). This radiological finding was not associated with trigeminal neuralgia, but sometimes (37.5%) with painless paresthesia of the V3 territory (4). Interestingly, when T2-weighted sequences were available (3,4) an ancient (non-enhancing) or recent (contrast-enhancing) linear pontine demyelinating lesion was always noted, in continuity with the emergence of the affected V<sup>th</sup> nerve. Some authors have suggested that such enhancement of the peripheral V<sup>th</sup> nerve in MS patients may reflect a demyelinating process specifically targeting the peripheral myelin (2,4). Indeed, the transition zone between central and peripheral myelin of the V<sup>th</sup> nerve is located less than 7 mm from the REZ (5); thus, an enhancement throughout the entire length of the cisternal V<sup>th</sup> nerve does reflect an involvement of the peripheral myelin. However, the constant presence of an adjacent intrapontine lesion, when T2-weighted images were analyzed, suggests that this involvement of peripheral myelin may correspond to a point-to-point extension of the inflammatory reaction, rather than a specific peripheral myelin demyelination process. This extension may be driven by a mechanical progression of edema along the cranial nerves, or by the propagation of inflammatory changes along the venous structures adjacent to the cranial nerves. The fact that some patients present with an enhancement of the peripheral V<sup>th</sup> nerve, while the adjacent central demyelinating lesion does not enhance, suggests that inflammatory changes may persist longer in the peripheral nerve than in the CNS (4)

A late occurrence of hypertrophy of the V<sup>th</sup> nerve, without contrast enhancement, in patients with long-lasting MS (3), may reflect an efficient remyelination process within the peripheral nerve. Interestingly, trigeminal enhancement is more frequent in secondary progressive MS patients, which could be explained by the greater dissemination and duration of the disease (4).

### **Conclusion:**

We report the second case of radiological involvement of cisternal peripheral III<sup>rd</sup> nerve in an MS patient. Radiological involvement of a cisternal cranial nerve in MS patients is exceptional for the III<sup>rd</sup> nerve, but has been reported more frequently for the V<sup>th</sup> nerve. Given the constant association with an adjacent central lesion on T2-weighted images, we suggest that such involvement of cisternal peripheral cranial nerve in MS reflect a local propagation of inflammatory changes, rather than a specific targeting of the peripheral myelin

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