

Hypertrophic olivary degeneration: unveiling the triangle of Guillain-Mollaret

Degeneração olivar hipertrófica: descobrindo o triângulo de Guillain-Mollaret

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A 28-year-old woman presented with hemi-dystonic movements on the left. Four years earlier, a ruptured vascular malformation caused a hemorrhagic stroke on the left cerebellar hemisphere (Figure 1B), evacuated by surgery. MRI revealed *right* hypertrophic olivary degeneration (HOD) (Figure 1A) and reduction of cerebellar-rubro pathways (Figure 2).

HOD is a trans-synaptic degeneration affecting the dentate-rubro-olivary pathway or Guillain-Mollaret's triangle¹. A lesion in any part of this synaptic pathway may lead to HOD (Figure 3), including stroke, cranioencephalic trauma, neurodegenerative diseases, inflammatory illnesses, among others^{2,3}. This unique synaptic collapse is associated to palatal myoclonus, parkinsonian features and dystonia^{1,2}.

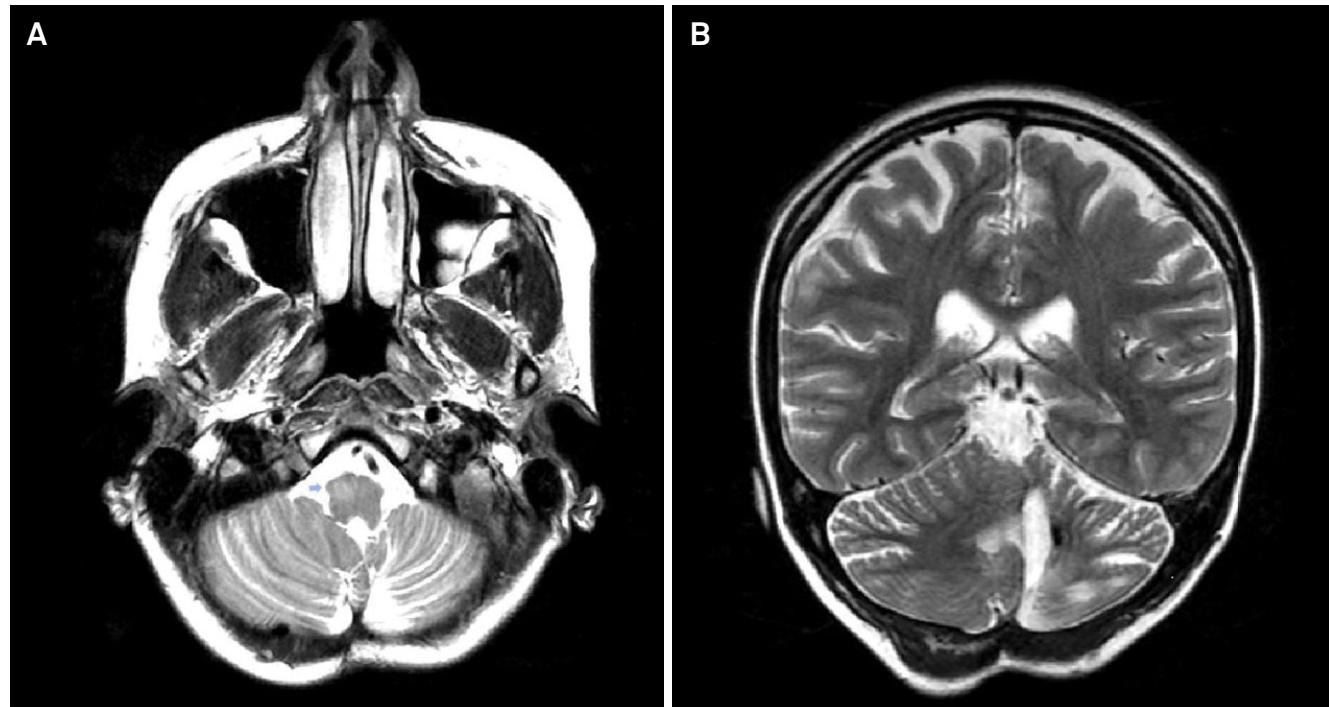


Figure 1. (A) Axial T2-weighted image showing enlargement and hyperintensity of the *right* olfactory nuclei, compatible to hypertrophic olfactory degeneration; (B) Coronal T2-weighted image displaying reduced *left* cerebellar hemisphere associated to hemosiderin.

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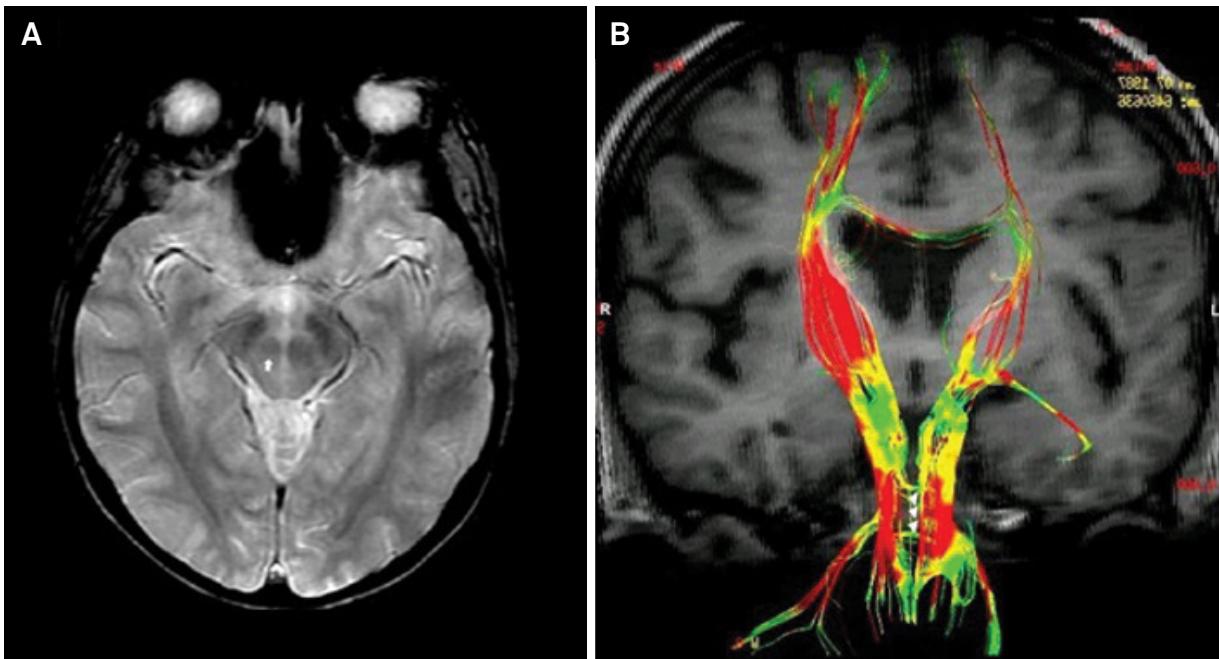


Figure 2. (A) Atrophy of right red nucleus (arrow) and substantia nigra; (B) Tractography image showing decreased volume of the right central tegmental tract (arrowheads).

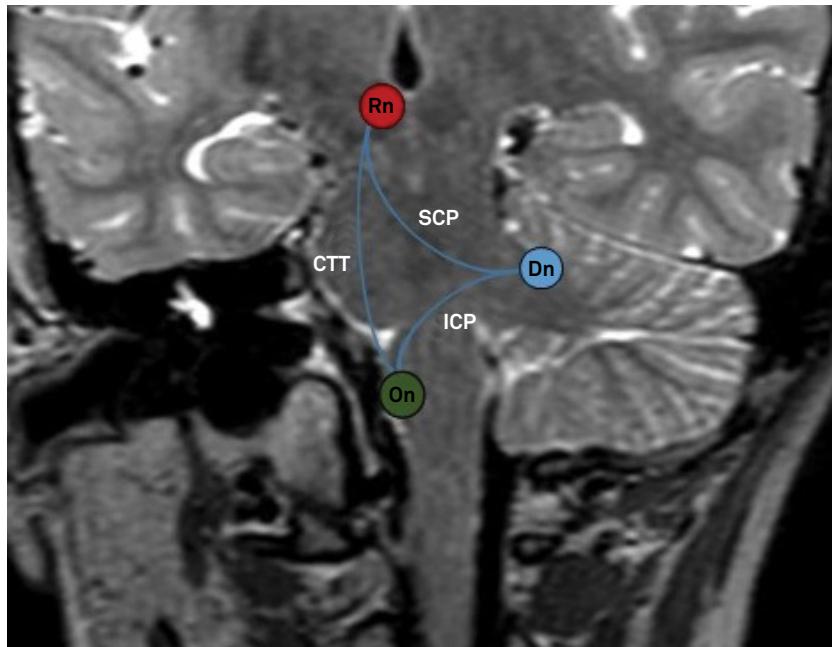


Figure 3. Anatomical view of the Triangle of Guillain-Mollaret. Initially, the axons from the dentate nucleus of cerebellum travel through the superior cerebellar peduncle (SCP) and decussate in the brachium conjunctivum to terminate into the contralateral red nucleus (Rn). Then, the central tegmental tract (CTT) leaves the red nucleus in the direction of the ipsilateral inferior olivary nuclei (On), situated downwardly at the medulla oblongata. At last, the inferior cerebellar peduncle (ICP) connects the ION to the opposite dentate nucleus (Dn), closing this complex pathway known as dentate-rubral-olivary circuitry.

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