LETTERS TO THE EDITOR

Regarding the famous triangle of Guillain-Mollaret*

Sobre el famoso «triángulo» de Mollaret

Dear Editor:

An article published in Neurología by Dr Sanchez-Hernandez et al.1 presents 2 cases of possible hypertrophic olivary degeneration secondary to a lesion of the upper cerebellar peduncle and central tegmental tract. These cases are similar to others reported previously.2 I would like to comment on the anatomical background they provide in the article.

It is true that the dentate nucleus projects to the red nucleus and projections extend from that location to other structures of the brainstem, including the inferior olivary nucleus and thalamus. Nevertheless, the main dentato-olivary pathway does not pass through the red nucleus. It is a direct pathway that passes by the red nucleus without forming synapses there (Figure 17.24 and Figure 1).3,4 This explains hypertrophic degeneration of the olivary nucleus, one of the best examples of trans-synaptic degeneration in the human brain. If the main dentato-olivary pathway were to form a synopsis in the red nucleus, any trans-synaptic degeneration would take place at that location and not the olivary nucleus, which would mainly receive a second projection. Furthermore, this scenario would not respect the precise topographical relationship existing between the dentate nucleus and the olivary nucleus, which I will explain below.

The lower side of the famous triangle of Guillain-Mollaret, described by Sanchez-Hernandez et al. as a direct olivo-dentate pathway, does not exist, as I have already stated in another publication.5 The main projections from the olivary nucleus pass through the inferior cerebellar peduncle and ascend as climbing fibres (described by Cajal) to the Purkinje cell dendrites but not to the dentate nucleus. It is true that some textbooks include drawings of abundant direct olivo-dentate fibres. While this may accurately describe the brains of certain animal species, it does not correspond to the human brain in which these direct fibres do not exist. The olivo-dentate connection is established by means of collateral projections of climbing fibres and these collateral projections are scarce (“olivocerebellar fibres only branch very sparingly”).6

In any case, the hypothetical olivo-dentate connection is irrelevant to phenomena of interest in clinical medicine, such as palate tremor, since injury to olivary projections (in the inferior cerebellar peduncle) cannot cause hypertrophic olivary degeneration whether directly or indirectly.

Given that these authors not only analyse neuroimaging in hypertrophic olivary degeneration but also elaborate on the neuroanatomy and neuropathology of the dentato-olivary pathway, I would like to have read the name of my professor Jean Lapresle among the references. Lapresle has probably contributed more to this topic than any other author by describing the precise topographical connections between these two structures and the mesencephalic tract of the dentato-olivary pathway adjacent to the red nucleus.7–10

References


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T1 hyperintensity in the pulvinar: A pathognomonic sign of Fabry disease?☆

Hiperintensidad pulvinar en T1: ¿un signo patognomónico de enfermedad de Fabry?

Dear Editor:

Fabry disease is an X-linked disorder of lysosomal metabolism that causes glycosphingolipid deposits in different tissues, with vascular endothelium being particularly susceptible. Clinical manifestations vary depending on the patient’s age at onset; they include skin lesions, acroparesthesia, pain episodes, anhidrosis, corneal opacity, hearing loss, and others. However, its vascular complications in the kidneys, heart, and brain are severe, especially in late-onset cases or in patients with longer histories of the disease. Although other types of neurological manifestations may be present, cerebrovascular problems are particularly frequent and serious.1,2 Doctors consider Fabry disease in the differential diagnosis of strokes of undetermined causes, and these strokes are most commonly linked to the vertebrobasilar territory.

Early diagnosis of this disease is important, given the availability and usage recommendations for enzyme replacement therapy with recombinant acid alpha-glucosidase.3 For example, researchers have described that hyperintensities in both pulvinar nuclei in a T1-weighted MRI sequence may be a pathognomonic sign for Fabry disease.4

We present the case of a patient with ischaemic stroke and bilateral pulvinar hyperintensities whose enzyme analysis was negative for Fabry disease. This 64-year-old man with hypertension and dyslipidaemia experienced sudden-onset dizziness and instability together with loss of lower limb strength and impaired enunciation. Neurological examination observed right hemiparesis which abated over the next few hours. Cranial CT revealed hyperintensities in both pulvinar nuclei (Fig. 1). A T2-weighted MRI also showed a hyperintense lesion in the left hemi-pons with restricted diffusion; findings are compatible with a recent infarct. In the T1-weighted sagittal sequence, we also observed hyperintensity restricted to both pulvinar nuclei (Fig. 2). Hyperintense lesions in the subcortical and periventricular white matter are also apparent in the T2-weighted sequence. There were no clinical signs or symptoms that would indicate Fabry disease, and the patient had no family history of the entity. Spectrofluorimetric determination of acid alpha-glucosidase in blood showed an activity level of 100%. We also ruled out other causes of calcification in the basal ganglia by measuring calcium, parathyroid hormone, glucose, and ammonia; HIV serology was also tested. It is therefore possible to state that pulvinar hyperintensity in this case may be secondary to small-vessel impairment due to arterial hypertension. Furthermore, pulvinar hyperintensity was secondary to calcifications at this location according to CT and MRI; similar findings have also been demonstrated in studies of Fabry disease.

To the best of our knowledge, this is the first reported case in which T1-weighted images of pulvinar hyperintensity are not linked to Fabry disease. Two other case studies have questioned whether the sign is really pathognomonic; in both, however, hyperintensity was not limited to the pulvinar nucleus and also affected the lenticular nucleus.5 The frequency of this radiological sign in patient series with Fabry disease4,6 has been estimated at 25% in men, but exceptionally rare in women. For this reason, theories suggest that this pattern could be related to the lower enzymatic activity in men, which would result in

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Figure 1   Brain CT. Hyperdensities can be seen in both pulvinar nuclei.