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ORIGINAL ARTICLE

Magnetic resonance imaging of hypertrophic olivary degeneration☆

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KEYWORDS
Magnetic resonance imaging;
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Olivary nucleus;
Red nucleus;
Cerebellar nucleus;
Medulla oblongata;
Cerebellum

Abstract
Objective: To review the pathophysiologic mechanisms involved in hypertrophic olivary degeneration, with attention to epidemiological and clinical aspects and especially to imaging findings.
Material and methods: We reviewed 5 patients diagnosed with hypertrophic olivary degeneration at our center from 2010 through 2013, analyzing relevant clinical, epidemiological, and radiological findings.
Results: In all cases, a hyperintensity was seen in the inferior olivary nuclei in FLAIR and T2-weighted sequences. No signal alterations were seen on T1-weighted sequences, and no enhancement was seen after intravenous injection of contrast material. In the cases studied by diffusion-weighted imaging, no significant alterations were seen in these sequences. Olivary hypertrophy was seen in all patients except in one, in whom presumably not enough time had elapsed for hypertrophy to occur. The alterations were bilateral in two of the five cases. Only one case exhibited the typical clinical manifestations.
Conclusion: Given that patients may not present clinical manifestations that can be attributed to hypertrophic olivary degeneration, it is important to recognize the characteristic radiologic signs of this entity.

PALABRAS CLAVE
Resonancia magnética;
Tronco cerebral;
Degeneración olivar hipertrófica;

Imagen por resonancia magnética en la degeneración olivar hipertrófica

Resumen
Objetivo: Repasar los mecanismos fisiopatológicos de la degeneración olivar hipertrófica, prestando atención a los aspectos epidemiológicos y clínicos, y sobre todo a los hallazgos de imagen.

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Núcleo olivar;
Núcleo rojo;
Núcleo cerebeloso;
Bulbo raquídeo;
Cerebelo

Material y métodos: Se revisaron 5 pacientes diagnosticados de degeneración olivar hipertrófica en nuestro centro entre los años 2010 y 2013, analizando los aspectos clínicos, epidemiológicos y radiológicos relevantes.
Resultados: En todos los casos se vio una hiperintensidad en los núcleos olivares inferiores en las secuencias FLAIR y T2. Las secuencias potenciadas en T1 no mostraron alteraciones de señal ni tampoco se observó realce tras inyectar contraste intravenoso. En los casos en los que se realizó una secuencia de difusión, no hubo alteraciones significativas. Salvo en un paciente, en el que presumiblemente no había pasado el tiempo necesario, en todos los restantes se vio una hipertrofia olivar. Las alteraciones fueron bilaterales en dos de los cinco individuos. En solo un caso las manifestaciones clínicas fueron típicas.
Conclusión: Dado que los pacientes pueden no presentar manifestaciones clínicas atribuibles a la degeneración olivar hipertrófica, resulta importante reconocer los signos radiológicos característicos.

Introduction

Hypertrophic olivary degeneration (HOD) is an uncommon pathological entity characterized by trans-synaptic degeneration secondary to lesions in the dento-rubro-olivary tract or ''the Guillain–Mollaret triangle'' discovered in 1931 by Guillain and Mollaret.[1] The expression ''trans-synaptic neuronal degeneration'' makes reference to the alteration of a group of neurons when a destructive process interrupts the majority of its afferent impulses.[2] The degeneration can occur in other locations yet the HOD has been considered a unique morphological type where hypertrophy of the degenerated neurons occurs.[3] The lesions in this tract are caused most often by diseases of vascular origin (ischemic or hemorrhagic) but it can also be the cause of a trauma or a tumor or due to surgical manipulation, infections, demyelinating or degenerative diseases.[4] A percentage of cases, which in some series is around 40%, is of unknown cause or without any visible lesions. We presented a series of five patients with hypertrophic olivary degeneration with special emphasis on the clinical and epidemiological aspects and magnetic resonance (MR) findings.

Materials and methods

The dento-rubro-olivary tract (Fig. 1) connects the red nucleus of the mesencephalon, the inferior olivary nucleus of the medulla oblongata and the dentate contralateral nucleus of the cerebellum. Fibers originating in the red nucleus descend through the central tegmental tract until they reach the inferior ipsilateral olivary nucleus. In turn, the olive projects fibers to the contralateral dentate nucleus through the inferior cerebellar peduncle crossing the midline at the height of the inferior olivary nucleus. To complete the triangle, efferent fibers of the dentate nucleus ascend through the superior cerebellar peduncle and they decussate until they synapse in the contralateral red nucleus.[4–15] HOD is due to lesions that affect the central tegmental tract or the dento-rubral tract of the Guillain–Mollaret triangle which will cause olivary disconnection.[16] We have reviewed the relevant clinical, epidemiological and radiological aspects of 5 patients diagnosed with HOD at our hospital from 2010 through 2013. All of them were examined through an MRI, two for the control of the underlying disease (ependymoma and pontine hemorrhage) and three because they showed clinical syndromes (cerebellar syndrome and cognitive deterioration). The studies were conducted in a Siemens MAGNETOM Symphony Maestro Class 1,5T (Siemens Medical Systems, Erlangen, Germany). In four patients FLAIR axial, axial T2-weighted sequences were acquired and axial T1-weighted sequences with and without intravenous contrast, and diffusion with apparent diffusion coefficient map.

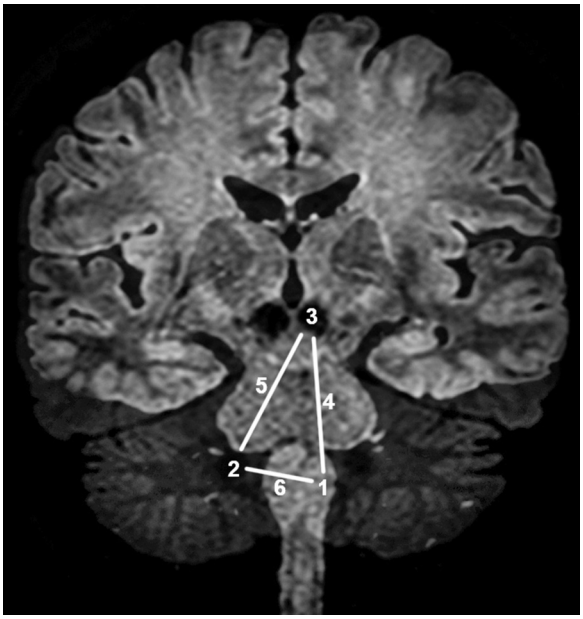

Figure 1 Sketch of the dento-rubro-olivary tract or ''the Guillain–Mollaret triangle''. 1: olivary nucleus. 2: dentate nucleus. 3: red nucleus. 4: central tegmental tract. 5: dentorubral tract. 6: dento-olivary tract.