CASE REPORT

BILATERAL HYPERTROPHIC OLIVARY DEGENERATION SECONDARY TO ISCHEMIA FOLLOWING ENDOVASCULAR TREATMENT OF CEREBRAL ANEURYSM: A CASE REPORT

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ABSTRACT

Hypertrophic olivary degeneration (HOD) is a degenerative disorder of the inferior olivary nucleus (ION) that occurs after damage to the dentatorubroolivary pathway (DROP). Damage to the DROP causes hypertrophy and enlargement of the inferior olivary nuclei (ION) contrary to the atrophy observed in the other parts of central nervous system. Focal lesions that may lead to HOD include ischemia, hemorrhage, trauma, and cavernous hemangioma and it also may mimic diseases including tumors and demyelinating processes. We here in present our experience with a case of bilateral HOD, which was a result of a thromboembolic complication during endovascular aneurysm embolization.

Key Words: Hypertrophic olivary degeneration, dentatorubroolivary pathway, thromboembolic, endovascular.

ÖZET

Hipertrofik olivar dejenerasyon (HOD) inferior olivar nukleusun dentatorubro-olivar yolakta (DROP) oluşan herhangi bir hasara yant olarak oluşan trans-sinaptik dejenerasyon türüdür. Santral sinir sisteminde bir anatominin bollüğün dejenerasyonu sekonder gelişen nöron kaybı ve glial hücrelerinde göğüsman aksine DROP yolakında transnöronal dejenerasyon sonucu hipertrofi gelişmektedir. HOD, tümör ve demyelinizan hastalıklar dahil pek çok patolojisi taklit edebilen, kanama, travma ve kavernöz hemanjioma bağlı gelişen, göreceli olarak nadir rastlanan bir durumdur. Endovasküler anevrizma embolizasyonu sırasında tromboembolik kompleksasyon sonucu gelişen, bilateral HOD olgusuya deneyimimizi paylaşıma amaçlıdik.

Anahtar Sözcükler: Hipertrofik olivar dejenerasyon, dentatorubro-olivar yolak, tromboembolic, endovasküler.

INTRODUCTION

Hypertrophic olivary degeneration (HOD) is a degenerative disorder of the inferior olivary nucleus (ION) that occurs after damage to the dentatorubroolivary pathway (DROP) (also called the Guillain–Mollaret triangle, or GMT). Damage to the DROP causes hypertrophy and enlargement of the inferior olivary nuclei (ION) contrary to the atrophy observed in the other parts of central nervous system. The magnetic resonance imaging features shows typical radiological temporal course that correlates with histopathological findings (1).

Bilateral involvement is extremely rare. Clinically, the patient may present with palatal tremor, dentorubral tremor and features of cerebellar/brainstem dysfunction. (2,3) Focal lesions that may lead to HOD include ischemia, hemorrhage, trauma, and cavernous hemangioma and it also may mimic diseases including tumors and demyelinating processes. We here in present our experience with a case of bilateral HOD, which was a result of a thromboembolic complication during endovascular aneurysm embolization.
CASE

A 60-year-old male was admitted to our emergency department with sudden onset of headache and blurred vision. His medical history was unremarkable. Non-enhanced brain tomography revealed an aneurysmatic dilatation located in right ambient cistern. Digital subtraction angiography showed a wide-necked right superior cerebellar artery aneurysm. The patient was treated with stent-assisted coil embolization (Figure I). The procedure was completed and the patient recovered with left facial paralysis and left hemiparesis. Diffusion-weighted imaging immediately after procedure showed an acute ischemia in the posterior aspect of right cerebral peduncle and superior cerebellar peduncle, caused by thromboembolic occlusion of a perforating artery arising from stented proximal right posterior cerebral artery (Figure II). Two months after initial insult, in his routine control visits, T2-weighted MR images showed enlargement and increased signal intensity in both olivary nuclei, consistent with bilateral HOD (Figure III). Still his clinical examination was unremarkable, except for dizziness responsive to symptomatic treatment.

DISCUSSION

The anatomical triangle named after Guillain and Mollaret, first described in 1931, consists of the ipsilateral red nucleus, the inferior olivary nucleus and the contralateral dentate nucleus (4). Anatomically the edges of the triangle include the connection between superior cerebellar peduncle to ipsilateral red and the contralateral dentate nuclei and the other connection of inferior cerebellar peduncle to contralateral dentate and the ipsilateral and inferior olivary nuclei. While hypertrophic olivary degeneration can be caused by any lesion involving the aforementioned structures, it is typically seen with focal lesions that disrupt the afferent pathways to the olive (dentatorubral and rubro-olivary pathways).

HOD is almost always unilateral; however, rare bilateral cases have been reported. Midline lesions or lesions in the brachium conjunctivum (superior cerebellar peduncle), finally interrupting decussation of the DROP, can result in bilateral HOD. In CNS, transneuronal degeneration associated with atrophy of the targeted structure is a common response to a confined lesion. Still transneuronal degeneration resulting in...
hypertrophy of the targeted region is unique to the inferior olivary nucleus (5). Pathologic changes including vacuolar degeneration, neuronal enlargement, astrocyte hypertrophy, demyelination, and gliosis are reflected in the typical imaging appearance of HOD, with an increase in signal on T2 and proton density weighted images and an increase in size of the olivary nucleus (6).

Depending on the time interval between the event and the scanning procedure, the size of the hypertrophic olivary nucleus is variable, with a normal size in the acute stage. Olivary hypertrophy typically develops around six months after the event and resolves after three to four years. Radiologically, an increased signal on T2 and proton density images confined to the olivary nucleus or nuclei (with or without ION enlargement) with lack of contrast enhancement or diffusion restriction typically appears early (around one month after the initial lesion) and persists infinitely (1, 7). Three stages of hypertrophic olivary degeneration can be seen on T2 weighted MR WMR images. In the first stage occurring within the first 6 months, only hyperintensity of the inferior olivary nucleus is seen. The second phase lasts from 6 months up till 3 to 4 years, and is characterized by an increase in size and intensity with non-visualization of the pre- and post-olivary sulci. The third stage begins with the resolution of the hypertrophy with persistence of the hyperintense signal in the olivary nucleus, which may last indefinitely (7).

The clinical appearance of the hallmark symptoms which consist of palatal myoclonus and other involuntary movements (dentatorubral tremor, ocular myoclonus and cerebellar or brainstem dysfunction) persists despite the resolution of the imaging characteristics of HOD. These symptoms presumably reflect loss of inhibitory control that is transmitted through dentat-rubral pathway (8). The differential diagnosis of signal hyperintensity n T2 weighted images within the pontomedullary region includes tumors, demyelinating lesions, infarction, and inflammatory processes (tuberculosis, sarcoidosis, or encephalitis). The lack of contrast enhancement, however, is contrary to many tumorous entities or an infectious origin, while the additional enlargement of the olivary nucleus is contrary to chronic stages of infarction or multiple sclerosis. MRI fiber tractography also can demonstrate disruption of the triangle of Guillain-Mollaret and can be useful in the diagnosis of HOD when findings on conventional MRI imaging are equivocal (9). Dincer et al., apart from documenting disruptions in the triangle of Guillain-Mollaret in HOD, showed increase in radial and axial diffusivity in the inferior olives, representing demyelination and neuronal hypertrophy, respectively (10).

In conclusion, diagnosis of HOD should be considered in a patient with dento-rubroolivary pathway lesions. The bilateral HOD is extremely rare and it is also important to differentiate between HOD and other medullary lesions. Familiarity with the clinical and radiological findings, which are very characteristic, is crucial and would avoid unnecessary diagnostic tests.

REFERENCES