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Hypertrophic Olivary Degeneration: A Case Report and Review of Literature

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Abstract

Hypertrophic Olivary Degeneration (HOD) is a type of transsynaptic degeneration caused by disruption of the dentato-rubral olivary pathway, Most of the neurodegenerative diseases are secondary to midbrain, pontine or cerebellar lesions. The main clinical manifestations were persistent dizziness, palatine myoclonus, unstable walking, and central nystagmus and cerebellar ataxia were observed on physical examination. Typical radiographic findings were enlargement of the medulla oblongata and abnormal signal in the olivary nucleus.

Keywords

Hypertrophic Olivary Degeneration; HOD; Case Report; MRI.

1. Introduction

HOD, a rare cross-synaptic neuronal degenerative disease, is often secondary to cerebellar dentate nucleus, midbrain red nucleus and olivary oblongata ring(denta-torubro-olivary pathway, DROP). The lesions (including hemorrhage, infarction, trauma, inflammation or tumor) in the midbrain, tegmental pons, superior crus and cerebellum may cause the destruction of the above circuits, leading to anterograde vacuoloid degeneration of the submedulla oblongata distant from the primary site, which is morphologic as focal hypertrophy of the inferior olive nucleus.This disease is relatively rare in clinical practice and is often misdiagnosed as cerebral infarction, demyelination or tumor.

2. Presentation of case

A 53-year-old woman with no major past medical history was referred to our department for a chief complaint of dizzinessing for more than 8 months and aggravation accompanied by blurred vision for 2 months. The patient suddenly experienced dizziness at work 8 months ago and fainted to the ground. She went to a local hospital for CT examination which indicated brain stem hemorrhage,and then discharged after conservative treatment.

After discharged to home, the patient continued to feel dizzy and drowsy, without headache, nausea and vomiting, see things rotating. patients with dizziness is aggravating especially standing up 2 months ago, decubitus declined slightly, activity significantly restricted, can't walk normally, daily life cannot provide for oneself, gradually appeared around double vision, with facial ministry, the tip of the tongue, fingertips numbness of limbs, and gradually increase.

A history of high blood pressure more than 3 years, the highest blood pressure to 170/110 mmHg, no regular monitoring of blood pressure and oral antihypertensive drugs, denied history of diabetes and coronary heart disease. Physical examination at admission :T:36.5℃,BP 130/70 mmHg, no yellow staining on the skin of the whole body, no swelling of superficial lymph nodes of the whole body, clear respiratory sound in both lungs, no rale of dry or wet was

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heard heart rate is 80 times/min, the rhythm is uniform, no murmur is heard on auscultation of each valve, the abdomen is flat, no swelling is touched under the liver, spleen and costal, no tenderness, rebelling pain, no deformity of the spine and limbs, no edema of the lower limbs.

Neurological examination shows that numbness of the tip of the tongue in the face,both sides of the pupils are equally large and equally round, with a diameter of about 3mm. They are sensitive to light reflection, and the eyes can move freely. The visual objects of both eyes are double, and the frontal lines of both sides of the nystagmus are symmetrical.Numbness in the fingertips of the limbs, normal muscle strength and muscle tension, finger-nose test,heel-knee tibial test,eye closed and difficulty standing sign (-), symmetrically felt sensation in the limbs, symptomatic symptom of bilateral tendon reflex, suspicious positive Pap sign on the left side, negative Pap sign on the right side, soft neck, bilateral Kirschner sign and Brinchner sign (-).

MRI suggested: 1. A kind of circular abnormal signal shadow was seen on the right side of the medulla oblongata with a clear boundary and a size of about 0.8 x 0.7cm. T1W1 showed low signal (Figure. 1A), T2W2 showed slightly high signal (Figure. 1B). No enhancement was observed on enhanced scan. MRS indicated that: compared with the normal cerebellar parenchyma on the right side, the medulla oblongata lesions had a decreased NAA peak, Cr peak, CH peak, NAA/ CH peak, and CH/Cr peak (Figure. 1C). DTI showed that the bilateral cerebral hemisphere fiber tracts were roughly symmetrical, no obvious fiber destruction was observed (Figure.1D)

After neuronutrition was given and circulation improved, the patient complained of improvement of symptoms and was discharged.

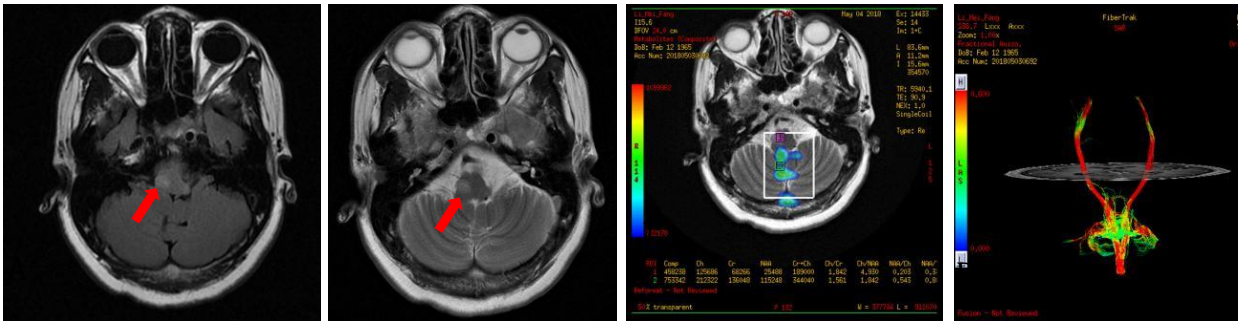

Figure 1. (A) MRI T1W1 Scan showed low signal (red arrowed), (B) T2W2 Scan sshowed slightly high signal (red arrowed), (C) MRS showed a decreased NAA peak, Cr peak, CH peak, NAA/ CH peak, and CH/Cr peak, (D)DTI showed that no obvious fiber destruction was observed.

3. Discussion

HOD was first reported by German doctor Oppenheim in 1887 [1], who called it a special "transsynaptic degeneration", that is, the damage of inferior neurons causes changes in the number, structure and function of superior neurons, leading to local hypertrophy of inferior olivary nucleus rather than atrophy. In 1931, Guillain and Mollaret pointed out that HOD was related to the damage of the DROP (denta-torubro-olivary pathway) pathway, and made a detailed explanation of this pathway for the first time [2]. That's why the loop is also known as the Guillain-Mollaret Triangle (GMT).

The main pathological manifestations of HOD include [3]: vacuolar degeneration of neuronal cells in the inferior olivary nucleus, proliferation of astrocytes and glia to varying degrees, and no significant increase in the number of neurons.

HOD may be ipsilateral, contralateral, or bilateral to the primary lesion. Lesions involving one side of the red nucleus or central tegmental tract can be secondary to ipsilateral HOD, while

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lesions involving one side of the dentate nucleus or superior crus of the cerebellar can lead to contralateral HOD, and bilateral HOD can be secondary to bilateral lesions in these sites, and bilateral HOD has also been reported as secondary to unilateral lesions [4].

MRI examination is specific for the diagnosis of HOD, showing high or slightly high signal intensity on T2WI, T1WI, etc., on unilateral or bilateral inferior olivary nucleus ventrolateral medulla oblongata. Slightly lower signal, FLAIR, DWI and ADC show equal or slightly higher signal, and there is no abnormality in enhanced scanning. In addition to T2WI, the other MRI terms showed multiple signal changes. The hypertrophy of the inferior olivine nucleus is characterized by regular shape, clear boundary and enlarged convex shape [5].

Different drugs such as clonazepam, levodopa and dopaminergic drugs have been reported to relieve symptoms to varying degrees. Levetracetam is a novel antiepileptic drug that modulates the dopaminergic system and has been shown to reduce tremor, dyskinesia, and myoclonus. Deep brain stimulation has become the best option for patients with disabling tremor, especially for those who are resistant to drug therapy [6].

4. Conclusion

In summary, HOD is a rare transsynaptic degeneration secondary to the GMT, with specific site of onset and characteristic MRI findings. To be familiar with the anatomy of GMT triangle and the projection relationship of nerve, to know the development pattern of HOD and to be familiar with its classical imaging examination are helpful to improve the understanding of this disease.

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