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DETERIORATION OF PALATAL MYOCLONUS AFTER ACUTE THALAMIC HEMORRHAGE

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Abstract

Background: Palatal myoclonus (PM) is the hallmark of hypertrophic olivary degeneration (HOD); however, little is known regarding the association of thalamic lesions and PM. Case presentation: Here, we report a case of deteriorative PM after an acute small ventrolateral thalamic hemorrhage in a female Chinese patient with HOD. The sudden and severe deterioration of PM was preceded by at least 10 days of an occasionally occurring PM, which was related to an acute cerebellar hemorrhage 8 months earlier. A computed tomography scan upon admission showed a small intracerebral hematoma in the left ventrolateral thalamus, and a magnetic resonance imaging scan revealed the typical signs of HOD as well as a remote lesion in the dentate nucleus. Symptoms of PM were controlled by carbamazepine and clonazepam. Conclusion: These findings indicated that the damaged dentatothalamic tract might be due to a unique pathogenic mechanism involving a lesion of the ventrolateral thalamus and Guillain-Mollaret triangle.

Keywords

• Palatal myoclonus • Hypertrophic olivary degeneration (HOD) • Thalamic hemorrhage • Magnetic resonance imaging (MRI)

Introduction

Hypertrophic olivary degeneration (HOD) is a degenerative disorder that occurs due to lesions in the dento-rubro-olivary pathway (also known as the triangle of Guillain and Mollaret). It is considered unique amongst the types of degeneration because the affected inferior olivary neurons are enlarged rather than atrophic. The hallmark of this disease is palatal myoclonus (PM) characterized by a rhythmic involuntary jerking movement of the soft palate [1]. Conventional therapy of PM involves the administration of clonazepam and sodium valproate [2]. A correlation between PM and thalamic lesions has rarely been reported. In this case report, we summarize a case of HOD with deteriorative PM after an acute mild ventrolateral thalamic hemorrhage which was controlled by a combination of clonazepam and carbamazepine (a member of the class of narrow spectrum antiepileptic drugs, which are seldom used to treat PM). This case may partly confirm the hypothesis that PM can be associated with a lesion of the ventrolateral thalamus (aside from those involving the Guillain-Mollaret triangle) through a unique underlying pathogenic mechanism. Furthermore, carbamazepine may be effective for special PM treatment.

Case

A 66-year-old female woke up with a severe palatal tremor and was admitted to our hospital just after 11 h a.m. The presence of an occasional palatomyoclonus had been noted for at least 10 days (without any discomfort, except that caused by appearance). Eight months prior to the occurrence of the palatal tremor, the patient had suffered an acute hypertensive hemorrhage involving the left cerebellum. The resulting neurological deficits had been almost completely alleviated. Neurological examination revealed that rhythmic PM was present with involuntary contractions occurring at a frequency of three or five cycles per second. An initial computed tomography (CT) scan revealed a small intracerebral hematoma in the left ventrolateral thalamus (Fig. 1). On the third day, the patient developed a slight involuntary jerking movement of the diaphragm, and a detailed magnetic resonance imaging (MRI) scan revealed the typical signs of HOD with isointensity in the enlarged right olivary nucleus on T1-weighted images and hyperintensity on the corresponding T2-weighted fluid-attenuated inversion recovery (FLAIR) images (Fig. 2A, B). On susceptibility-weighted images (SWI), the remote hemorrhage of the left dentate nucleus was more visible than that on the T2-weighted FLAIR images (Fig. 2C, D). The patient's PM symptoms were controlled with oral clonazepam (2 mg, two times a day) and carbamazepine (0.2 g, three times a day).

Discussion

HOD is usually described as a form of trans-synaptic degeneration, which occurs subsequent to a lesion involving the contralateral dentate nucleus of the cerebellum, the ipsilateral red nucleus, and the inferior olivary nucleus in the Guillain-Mollaret triangle. It is unique in that the the affected

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inferior olivary neurons are enlarged rather than atrophic [1]. The characteristic feature of this disease is PM, which is characterized by a rhythmic involuntary jerking movement of the soft palate, often involving the diaphragm and laryngeal muscles [1]. Without knowledge of this clinical pathological entity, clinical presentations of this patient could have been misinterpreted as a new ischemic event, a mass lesion, or a progressive neurodegenerative disorder affecting the cerebellar system [3].

Here, we describe a patient who suddenly suffered from PM following an acute small thalamic hemorrhage. Most commonly, lesions involving the dentato-olivary tract are the cause of PM. The development of a symptomatic palatal tremor (SPT) depends on the hyperactivity of olivary neurons initiated from inhibitory inputs, particularly until the peak of both SPT and inferior olivary hypertrophy occurs [4]. The past history of a left dentate nucleus hemorrhage may explain the existence of PM; however, the reason for

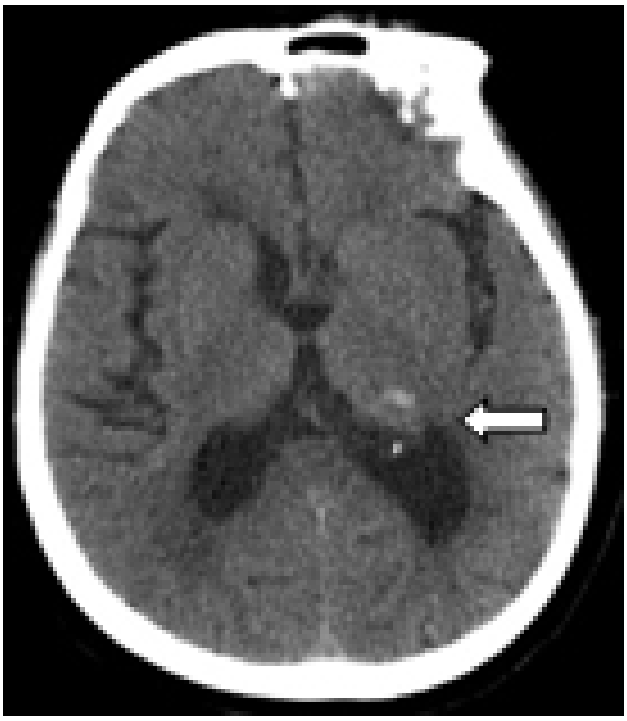

Figure 1. CT image showing a small intracerebral hematoma in the left ventrolateral thalamus.

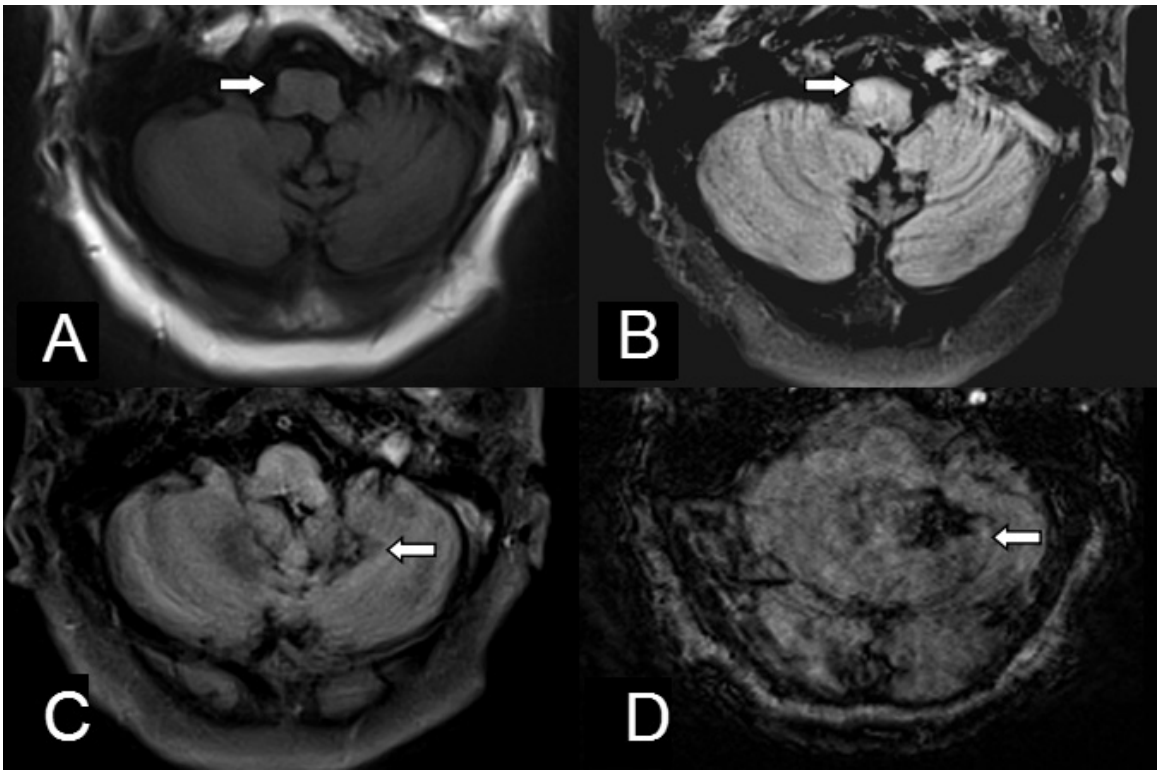

Figure 2. MRI revealed the typical signs of HOD. Isointensity in the enlarged right olivary nucleus is shown on the axial T1-weighted image (A), and hyperintensity is shown on the corresponding T2-weighted fluid-attenuated inversion recovery (FLAIR) image (B) (white arrow). The remote hemorrhage of the left dentate nucleus was better visible on susceptibility-weighted images (SWI) (D) than on the T2-weighted FLAIR image (C) (white arrow).

sudden deteriorative PM following an acute ventrolateral thalamic hemorrhage is unclear. Furthermore, the correlation between PM and thalamic lesions has rarely been reported. Cerrato *et al.* reported a case in which PM followed a lateral thalamic infarction in the absence of a brainstem or cerebellum lesion. They hypothesized that the influence of the lateral thalamus on the red nucleus was located in the upper mesencephalon [5]. Anatomically, the ventral oral posterior nucleus receives input from the dentate and red nuclei *via* the dentatothalamic tract and projects to the motor cortex [6]. Unfortunately, little research has been performed recently regarding thalamo-rubral fibers. Although the intracerebral hematoma in the left ventrolateral thalamus was small in our case as compared with that in the study by Cerrato *et al*. [5], the dentatothalamic tract might have been injured. Thus, our case may partially confirm the hypothesis that the unique pathogenic mechanism involves a lesion of the ventrolateral thalamus and the Guillain-Mollaret triangle. It is plausible that the dentatothalamic tract fibers may inhibit hyperactivity of the olivary neurons or control the feedback from the abnormal movement resulting from olivary neuron dysfunction.

Furthermore, as PM is a type of segmental myoclonus, clonazepam and sodium valproate are most frequently used to treat myoclonus [2]. Based on our experience treating PM, most of the symptoms can be controlled by clonazepam or a combination of clonazepam and sodium valproate; however, the deterioration of PM was not well controlled in our case. We thus tried to treat the patient with narrow spectrum antiepileptic drugs (carbamazepine combined with clonazepam), to control the severe PM, and they were effective.

Acute thalamic hemorrhage may be one of the major causes of PM deterioration; however, we could not confirm whether the dentatothalamic tract fibers were damaged anatomically. It has been shown that decreased central tegmental tract volume away from the site of the inferior olivary lesion in HOD can be detected by diffusion tensor imaging (DTI) and fiber tractography [7]. Unfortunately, our DTI scans failed to detect the lesion due to severe involuntary movements of the head arising from PM deterioration.

In conclusion, our findings indicate that a lesion of the ventrolateral thalamus, distinct from lesions in the Guillain-Mollaret triangle may be associated with a unique underlying pathogenic mechanism of PM.

Acknowledgments

Conflict of interest statement: The authors declare no conflict of interest. *Consent:*

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal. *Authors' contributions:* CJG carried out the study design, participated in the analysis and interpretation of data, and drafted the manuscript. KN participated in the design of the study and performed part of the acquisition of data. YDL also participated in the acquisition of data. ZW and ZHW participated in administrative, technical, and material support. DHM conceived of the study, critically revised the manuscript for important intellectual content, and supervised the study.

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