

Case Reports

Severe Bilateral Kinetic Tremor Due to Unilateral Midbrain Lesions

Majid Esmailzadeh^{1*}, Nesrin Uksul¹ & Joachim K. Krauss¹

¹Department of Neurosurgery, Hannover Medical School, Hannover, Germany

Abstract

Background: The dentatothalamic tract connects the dentate nucleus of the cerebellum with the contralateral thalamus and plays a major role in the pathogenesis of tremor. Unilateral lesions of the dentatothalamic pathway may affect its ipsilateral predecussational or its contralateral postdecussational course, which results either in ipsilateral or in contralateral tremor.

Case Report: Here, we present two patients with a unilateral midbrain lesion resulting in bilateral tremor. Both patients presented with severe kinetic tremor.

Discussion: The corresponding unilateral mesencephalic lesion affected both the ipsilateral predecussational and the ipsilateral postdecussational dentatothalamic tract originating from the contralateral dentate nucleus, which is very unusual and has not been outlined clearly before.

Keywords: Dentatothalamic tract, midbrain, superior cerebellar peduncle, tremor, brainstem

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*To whom correspondence should be addressed. E-mail: esmailzadeh.majid@mh-hannover.de

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Introduction

Midbrain lesions may cause a variety of different tremors depending on the exact site of the lesion within the midbrain.^{1–4} Patients with unilateral tremor may harbor an ipsilateral or a contralateral lesion according to its location along the dentatothalamic pathway, but in patients with bilateral tremor usually bilateral lesions are identified.^{3,5,6} It is exceedingly uncommon, however, that a unilateral lesion may cause bilateral tremor.

Case reports

Patient I

A 55-year-old female was admitted after she lost consciousness. Magnetic resonance imaging (MRI) demonstrated a right-sided mesencephalic hemorrhage with intraventricular bleeding. A ventricular drain was placed for cerebrospinal fluid diversion. The patient recovered slowly over the next few weeks. Four weeks after the hemorrhage she developed severe ataxia and postural instability. In addition, bilateral kinetic tremor of the upper extremities became evident (frequency 5 Hz,

variable amplitude up to 8 cm). The tremor was scored according to the Fahn–Tolosa–Marin Tremor Scale: at rest = 0, with posture = 3 (right) and 4 (left), and with action = 4.^{7,8} Repeat MRI showed partial resorption of the hemorrhage. Digital subtraction angiography showed an arteriovenous malformation (Spetzler–Martin grade III) within the tectum and tegmentum of the right mesencephalon extending to the superior cerebellar peduncle. The arteriovenous malformation was partially embolized and then resected via a midline suboccipital craniotomy.

Postoperatively there was no change in tremor severity. Medication with levodopa, gabapentin and tiapride had no effect on tremor severity, but clonazepam yielded mild amelioration; however, it had to be stopped because of sedation. MRI at 5 months after the hemorrhage demonstrated a circumscribed lesion in the dorsal aspect of the right mesencephalon extending from the superior cerebellar peduncle up to the mesencephalic-diencephalic transition zone (Figure 1A,B). The patient recovered slowly, although she did not become fully independent. The tremor improved spontaneously and no further medication was needed.

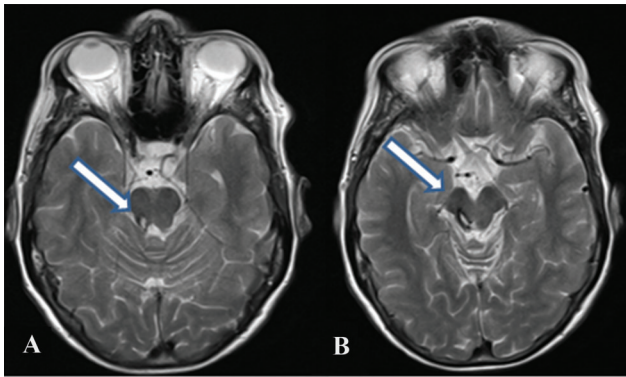


Figure 1. Axial T2-weighted Magnetic Resonance Imaging Scans of Patient 1 (A,B). A hypointense lesion with a hyperintense rim in the dorsal aspect of the right mesencephalon extending from the superior cerebellar peduncle to the mesencephalic–diencephalic transition zone.

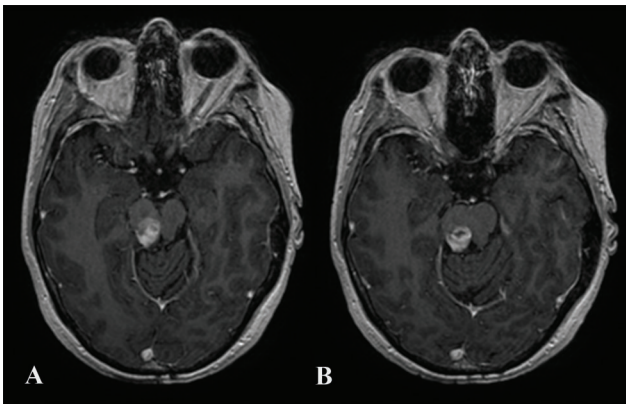


Figure 2. Axial T1-weighted Enhanced Magnetic Resonance Scans of Patient 2 (A,B). A tumor located in the right pontine and mesencephalic tectum and tegmentum.

Patient 2

A 41-year-old female presented with a 10-year history of slowly progressive left-sided hemiparesis, dystonia of her left hand, double vision, and ataxia. Initial MRI had demonstrated a small non-enhancing mesencephalic tumor, while recent studies showed an enhancing brainstem tumor located at the pontomesencephalic junction on the right side (Figure 2A,B). A stereotactic biopsy was performed, and histological examination revealed a glioblastoma World Health Organization grade IV. Despite combined radiochemotherapy there was a substantial increase in tumor volume. In parallel, the patient developed bilateral kinetic tremor of both upper extremities (frequency 4 Hz, amplitude 5–7 cm) as well as marked ataxia and dysarthria. Ratings on the Fahn–Tolosa–Marin Tremor Scale were the following: at rest = 0, with posture = 3, and with action = 4. Subtotal tumor resection was achieved by a telovelar approach via a midline suboccipital craniotomy.

Postoperatively, there was a mild transient increase in the right-sided tremor. During the next 2 years the situation stabilized under chemotherapy with temozolamid and bevacizumap. Subsequently,

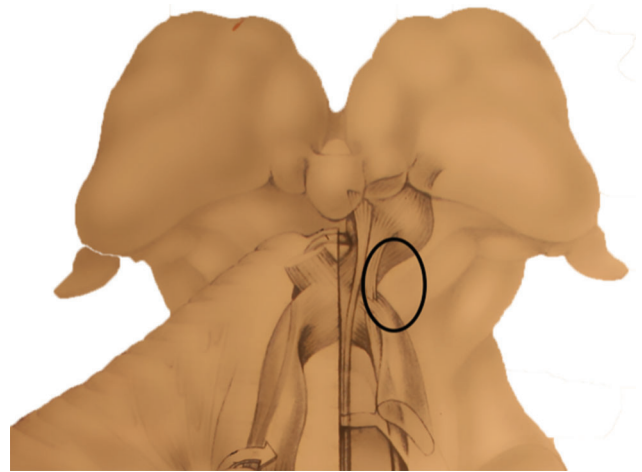


Figure 3. Schematic View of the Site of the Lesion. In both patients located at a strategic point close to the decussatio pedunculorum affecting the ipsilateral predecussational dentatothalamic pathway and the ipsilateral postdecussational dentatothalamic pathway originating from the contralateral dentate nucleus. (Modified from Figure 163, Nieuwenhuys et al., Springer, 1991 (13)).

however, there was an increase in dysarthria and ataxia, and the patient became bedridden. She also suffered from an increase in right-sided hemiparesis with parallel disappearance of the right-sided tremor. MRI demonstrated massive growth of the residual tumor. The patient succumbed within the next few weeks.

Discussion

Kinetic tremors as described in our patients have variously been labeled as midbrain tremor, rubral tremor, cerebellar tremor, or hyperkinesia volitionnelle.^{4,9} When such tremors are combined with a prominent tremor at rest, they may also be categorized as Holmes tremor according to a consensus statement.⁹

The anatomical correlate for kinetic tremor most frequently is a lesion of the dentatothalamic pathway along its course from the dentate nucleus to the contralateral thalamus. The dentatothalamic pathway leaves the cerebellum within the superior cerebellar peduncle entering the tectum of the midbrain and crosses to the contralateral side within the decussation of the superior cerebellar peduncles.¹⁰ While a predecussational lesion of the dentatothalamic pathway may cause tremor ipsilateral to the lesion, a postdecussational lesion would result in contralateral tremor.^{3,6}

There is a wide variety of disorders and lesions that may cause midbrain tremor, including multiple sclerosis, infection, stroke, trauma, tumor, cavernoma, or vascular malformation.^{1,4} The pathophysiology of kinetic tremor has not been fully clarified. Typically, such tremors develop with a variable delay of days or weeks after the lesion has occurred.^{9,11,12}

Bilateral kinetic tremor is more common in patients with multiple sclerosis in whom multiple lesions can be identified in the cerebellum or the midbrain.¹ A unilateral midbrain lesion may only cause bilateral tremor when it is located at a strategic point close to the decussation of the superior peduncles and if it has an appropriate size (Figure 3).^{10,13}

With regard to the published literature, such lesions seem to be exceedingly rare. Since Jakob and Huhn⁵ identified a vascular lesion affecting the mesencephalic tegmentum to be responsible for bilateral tremor during autopsy, this issue has received little attention.⁴

As demonstrated in our patients the prognosis of bilateral tremor associated with a unilateral lesion may depend both on the development of the lesion itself and also on the appearance of other neurological symptoms. While reduction in the volume of the lesion, such as increased resorption of the hemorrhage, may result in improvement of tremor, a marked increase in volume by tumor growth can also affect other systems such as the corticospinal tract and may result in reduction of tremor secondary to an increase in hemiparesis.

In conclusion, unilateral midbrain lesions of the cerebellar outflow may affect the ipsilateral predecussational and the ipsilateral post-decussational dentatothalamic tract originating from the contralateral dentate nucleus, which can result in bilateral kinetic tremor.

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