Case report

Holmes’ tremor caused by midbrain cavernoma

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Keywords: Holmes’ tremor; cavernoma; midbrain

Holmes’ tremor has been postulated as a syndrome attributed to those lesions that interrupt the dentate-thalamic and the nigrostriatal tracts thus causing both an action and a rest tremor. It may arise from various underlying structural disorders including multiple sclerosis, stroke, or tumors. So far, to our knowledge, few studies on Holmes’ tremor secondary to cavernoma have been reported. Here we report a case of disabling tremor, who harbored a cavernoma in the midbrain.

CASE REPORT

A 42-year-old woman was admitted to our department in 2005 because of 10 years’ tremor, which involved her left shoulder and upper limb. On attempting posture, the irregular low frequency tremor became grossly uncontrollable, and further exacerbated by any attempted movement. She had a partial right oculomotor palsy and a left hemiparesis, and had been misdiagnosed as Parkinsonism and treated with L-dopa. After the treatment, her symptoms continued to deteriorate. When she was referred to our department, she could not use her left arm owing to the refractory tremor, and MRI demonstrated a round lesion in the right ventral midbrain adjacent to the medial cerebral peduncle, which was characterized by a circumferential hypointensity rim (Fig. 1). Thus, an orbitozygomatic craniotomy was performed on the patient via a frontotemporal transsylvian route to access this medial portion of the midbrain. With a longitudinal pial incision between the frontopontine fibers and the pyramidal tracts in the peduncle, the cavernoma was totally removed en bloc (Fig. 2).

After the surgery, the tremor disappeared, and the muscle strength of the left lower extremity of the patient improved from degree 3 to degree 4. The strength of the upper extremity remained the same after the operation, and then increased to degree 4 during 1-year follow-up. However, the right oculomotor palsy has not been improved. Postoperative MRI showed a complete excision of the midbrain cavernoma (Fig. 3). Histological examination confirmed the diagnosis of cavernoma.

DISCUSSION

In 1904, Holmes described a tremor of fingers with a rotation at the wrist and elbow, which he named “rubral tremor”. He believed that the rubrospinal tract was involved in the generation of this tremor based on the observation of a patient with an organic lesion in the rubrospinal tract in the pons. Nowadays, it is generally accepted that Holmes’ tremor is a symptomatic tremor that caused by different lesions of the brainstem/cerebellum and thalamus. It is described as a combination of rest and postural and kinetic tremors. The tremor is often not as rhythmic as other tremors with a lower frequency varying from 2 to 5 Hz. It may be higher during active movement and disappear during sleep. The amplitude at rest may be small, but on attempting posture it become uncontrollable. Unlike most other forms of tremor, the proximal muscles may be more affected compared to the distal muscles. In addition, the tremor often associates with other signs of midbrain damage, such as hemiparesis and cranial nerve palsy.

Tremor is an uncommon symptom of cavernoma, since it is seldom found in the brainstem. In our case, the nidus was located at the anterior midbrain between the pyramidal tracts and the red nucleus, where is the site of substitute nucleus (Fig. 4). This anatomical location explains the predominantly parapyramidal symptom of the patient.

The treatment of Holmes’ tremor is difficult. Generally, pharmacotherapy is not effective. Despite radiosurgery has been used in the treatment of cavernoma in the delicate region, the incidence of radiation-induced complications has been reported getting higher. Some authors tried to treat Holmes’ tremor by stimulating the nucleus ventralis intermedius. However, the deep brain stimulation can only relieve the postural tremor in the distal segment of the contralateral upper extremity (distal tremor), and has no effect on the vigorous action tremor (proximal tremor). Theoretically, the neurological deficit impairment attributable to direct tumorous invasion can be alleviated by treating the primary lesion. With a surrounding gliotic layer, excision of the cavernoma without a damage to the vital structures of the brainstem is possible. The point is to obtain a fairly safe entry zone on the mesencephalon, which is the priority to achieve a successful resection of the midbrain.
Fig. 1. Preoperative MRI. A: Gd-enhanced T₁-weighted sagittal image demonstrating a popcorn-like rounded lesion in the anterior rostral brainstem. B: A rim of decreased signal intensity (arrows) at the periphery of a heterogeneous central signal is noticed on a T₂-weighted axial image.

Fig. 2. Intraoperative view. Through a frontal temporal transsylvian route, orbitozygomatic craniotomy was performed and the ventral right mesencephalon was well exposed. The safe entry zone into the midbrain is a small rectangular area outlined medially by the third cranial nerve (III) and the basilar artery (BA), inferiorly by the superior cerebellar artery (SCA), superiorly by the posterior cerebral artery (PCA), and laterally by the tentorium edge. This narrow but fairly safe window allows surgical access through the more medial part of the peduncle, sparing the motor tract, which occupies the intermediate 3/5 or so of the peduncle. Under a microscope, the optical nerve (II), the internal cerebral artery (ICA), and middle cerebral artery (MCA) medially, posterior communicating artery anteriorly (PCoA), trochlear nerve (IV) posteriorly, were also visualized. The PCA had been mobilized down from the peduncle before a longitudinal incision (bar) was created.

Fig. 3. Postoperative MRI. The sagittal (A) and axial (B) images showing a complete excision of the mesencephalic cavernoma.

Fig. 4. Sketch of the mesencephalon transect. The cavernoma (T) located at the medial peduncle surrounded by the corticospinal tract (CST), frontopontine tract (FPT), and substitute nucleus (SN). An incision between the CST and FPT was created. PTT: arietotemporal tract; III: the root filaments of the oculomotor nerve; 3: oculomotor nucleus; RN: red nucleus.

cavernoma. To access this medial peduncle area, an orbitozygomatic craniotomy via the frontotemporal transsylvian approach is recommended.

REFERENCES


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(Received November 21, 2006)

Edited by LUO Dan