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''Kissing'' Brainstem Cavernous Angiomas Removed Using a Trans-Fourth Ventricular Floor Approach

—Case Report—

Takahiro MURATA,[1] Tetsuyoshi HORIUCHI,[1] Yoshiki HANAOKA,[1]
Hisashi MURAOKA,[1] Tetsuya GOTO,[1] Kiyoshi ITO,[1]
Keiichi SAKAI,[1] and Kazuhiro HONGO[1]

[1]*Department of Neurosurgery, Shinshu University School of Medicine, Matsumoto, Nagano*

Abstract

A 39-year-old woman presented with a rare case of ''kissing'' brainstem cavernomas formed by separate lesions enlarging with simultaneous recurrent hemorrhages, which was successfully treated by staged resection using a trans-fourth ventricular floor approach. She had a familial history of cerebral cavernous angioma, and presented with a history of four episodes of sudden neurological deterioration. Magnetic resonance (MR) imaging obtained at each neurological event demonstrated two distinct brainstem cavernomas located in the pontine tegmentum and ventral part of the lower pons, both of which enlarged stepwise caused by simultaneous recurrent hemorrhages. Both cavernomas contacted and formed ''kissing'' lesions. She underwent midline suboccipital craniotomy in the prone position. The cavernoma in the pontine tegmentum was resected through a trans-fourth ventricular floor approach. Although ''kissing'' formation appeared on preoperative MR imaging, parenchyma was identified at the bottom of the removal cavity of the dorsal lesion, and resection was terminated. MR imaging following the first surgery revealed complete resection of the pontine tegmentum cavernoma and the ventral pontine cavernoma, which was located adjacent to the bottom of the removal cavity and aligned in same direction along the fourth ventricular floor approach. At 10 days after first surgery, she underwent the same procedure with the aid of neuronavigation to resect the ventral pontine cavernoma through the former removal cavity. This approach through the previous removal route, particularly for resection of ''kissing'' lesions which are difficult to access in the brainstem, is a technically feasible microsurgical procedure.

Key words: brainstem, cavernous angioma, kissing lesions, multiple cavernomas, trans-fourth ventricular floor approach

Introduction

Cavernoma, also known as cavernous malformation or cavernous angioma, is characterized by abnormally dilated vascular channels lined by a single layer of endothelium, and accounts for approximately 5–10% of all central nervous system vascular malformations.[4,8] Most cerebral cavernomas are located in the supratentorial region, and brainstem cavernomas are relatively rare, in 9–35% of cases.[4,8,13,22] The natural history of brainstem cavernoma is worse than that of cavernoma in other regions.[1,14] Surgical resection of brainstem cavernoma is required if the clinical symptoms deteriorate due to repeated bleeding.[1,12,14,20] However, surgical resection of a brainstem lesion often represents one of the most technically difficult interventional procedures because of the distinctive anatomical structure packed in a small region. We have performed surgery with preserved neuronal function and good postoperative performance status for patients with brainstem lesion using intraoperative electrophysiological mapping and monitoring.[7,9] The surgical approach for resection of brainstem cavernoma must be carefully planned on the basis of lesion location, accessible route, and minimal injury to the brainstem. Generally, access is recommended through the site at which bulging of the lesion or adjacent surface from the lesion is identified. If multiple symptomatic cavernomas are present in the brainstem, separate approaches or entry points may be selected for each cavernoma.

We describe a case of two symptomatic brainstem cavernomas, located in the pontine tegmentum and ventral part of the pons, which appeared as ''kissing'' lesions on magnetic resonance (MR) imaging. One of the ''kissing'' cavernomas was resected through a trans-fourth ventricular floor approach and the other was subsequently resected through the same route.

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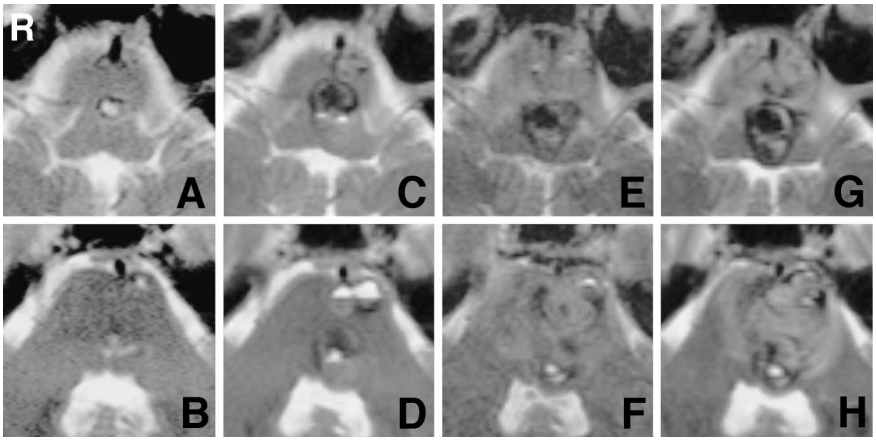

Fig. 1 T_2-weighted magnetic resonance images showing the chronological changes in the brainstem cavernomas. A, B: At the initial onset showing two small pontine cavernomas, located in the pontomedullary region and in the anterolateral part of the pons. C, D: At the second episode demonstrating both cavernomas had increased in size. E, F: At the third episode revealing both cavernomas had enlarged with perilesional edema. G, H: Axial images at the fourth event showing the cavernoma in the ventral pons with hemorrhage.

Case Report

A 39-year-old woman, with a familial history of cerebral cavernous angioma, presented with a history of four separate sudden neurological events. Her initial symptom at 6 years before admission in our institution was diplopia, which disappeared with conservative treatment. MR imaging demonstrated multiple cerebral cavernomas including two in the pons (Fig. 1A, B). During the 8 months before referral to our institution, three episodes of neurological deterioration occurred, consisting of diplopia, dysarthria, gait difficulty associated with moderate right hemiparesis as well as truncal ataxia, and right sensory dysesthesia. MR imaging obtained at each event revealed two brainstem cavernomas in the pons, both of which had enlarged stepwise with repeated hemorrhages (Fig. 1C–H). Simultaneous hemorrhages were thought to have occurred in both cavernomas at the second and third episodes, based on her symptoms and MR imaging findings.

On admission, she was well oriented and had normal cognitive function, but with diplopia due to left abducens nerve palsy and right sensory disturbance. Dysarthria, gait disturbance, truncal ataxia, and facial nerve palsy were denied. Preoperative MR imaging showed that the cavernoma in the pontine tegmentum bulged out to the fourth ventricular floor, and the cavernoma in the anterolateral part of the lower pons protruded to the left pontine surface (Fig. 2). The cavernomas appeared as ''kissing'' lesions. Treatment was primarily intended to resect the cavernomas in the pons via a trans-fourth ventricular floor approach. Staged resection was considered as location of the ventral pontine cavernoma was unclear intraoperatively.

She underwent midline suboccipital craniotomy in the prone position. The cerebellomedullary fissures were dissected and the floor of the fourth ventricle was exposed. The floor of the fourth ventricle bulged and the median sulcus was deviated to the right. With brainstem mapping, the pontine tegmentum cavernoma was entered from the infrafacial triangle[9] and was circumferentially dissected under intraoperative electrophysiological monitoring following evacuation of hematoma. Since transcranial motor evoked potential of right upper extremity temporarily attenuated during dissection at the deep lateral side of the lesion, we considered that the left corticospinal tract was displaced laterally by compression from the ventral pontine cavernoma. After resection of the pontine tegmentum cavernoma, parenchyma was identified at the bottom of the removal cavity and no apparent lesion was visible. As neuronavigation was not available in this case, removal was terminated although another ventral pontine cavernoma was present.

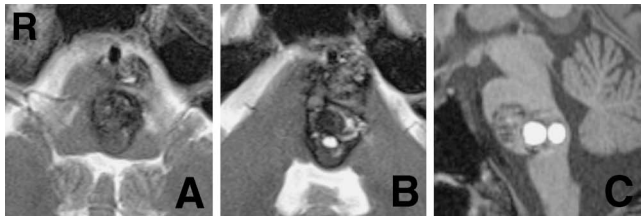

Fig. 2 A, B: Preoperative T_2-weighted magnetic resonance (MR) images showing that the cavernoma in the pontine tegmentum bulged out to the fourth ventricular floor, whereas the cavernoma in the ventral pons protruded to the left pontine surface, and formed ''kissing'' lesions. C: Sagittal T_1-weighted MR image using magnetization-prepared rapid acquisition gradient echo demonstrating both cavernomas in contact in the lower pons.

Postoperatively she had worsening right sensory disturbance but not right hemiparesis. MR imaging obtained 8 days later demonstrated complete resection of the cavernoma in the pontine tegmentum, and the remaining ventral pontine cavernoma located adjacent to the bottom of the removal cavity and situated in the same direction along the fourth ventricular floor approach (Fig. 3).

Consequently, resection of the remaining cavernoma was scheduled with neuronavigation to determine the entry point through the bottom of the removal cavity at 10 days after the first surgery. After the previous entry route on the floor of the fourth ventricle was identified, access to the ventral lesion was determined under neuronavigation guidance through the medial rostral side at the bottom of the previous removal cavity. The lesion was entered with a vertical incision for minimal invasion and was then dissected circumferentially under intraoperative monitor-

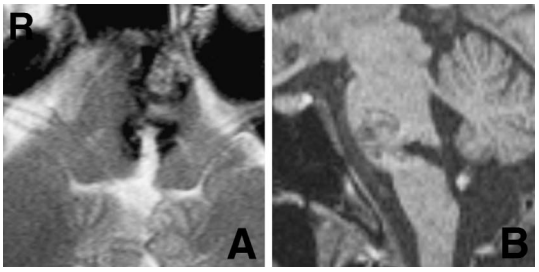

Fig. 3 A: Axial T_2-weighted magnetic resonance (MR) image at 8 days after first resection showing complete resection of the pontine tegmentum cavernoma and the remaining ventral pontine cavernoma. B: Sagittal T_1-weighted MR image using magnetization-prepared rapid acquisition gradient echo demonstrating that the remaining cavernoma adjacent to the rostral side in the removal cavity of resected cavernoma was aligned along the direction of the trans-fourth ventricular floor approach.

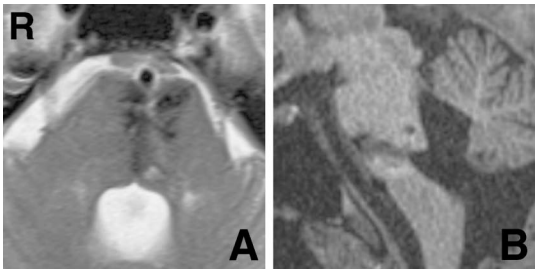

Fig. 4 A: Axial T_2-weighted magnetic resonance (MR) image showing complete resection of the ventral pontine cavernoma. B: Sagittal T_1-weighted MR image using magnetization-prepared rapid acquisition gradient echo revealing the enlarged entrance of the route on the floor of the fourth ventricle.

ing. The deepest part of the lesion was eventually removed, the pia mater of the ventral pontine surface was exposed, and total resection was achieved. She tolerated this procedure well.

Postoperatively she developed transient mild right hemiparesis, mild left facial nerve palsy, and dysphagia without tracheostomy. Follow-up MR imaging revealed complete resection of the cavernoma in the ventral pons and enlarged entrance of the route on the floor of the fourth ventricle (Fig. 4). The histological findings of both lesions were consistent with cavernoma. Her gait disturbance due to the right sensory disturbance remained and she was transferred for rehabilitation. She was ambulatory without a cane at 3 months after the second operation, and returned to her previous work.

Discussion

We describe a rare case of ''kissing'' cavernomas traversing anteroposteriorly in the pons, which was successfully treated by staged resection through a trans-fourth ventricular floor approach. Such ''kissing'' pontine cavernomas and the approach through the previous removal route via the trans-fourth ventricular floor are extremely unusual.

Cerebral cavernoma can occur in sporadic or familial forms, the latter associated with inherited autosomal dominant disorder and often characterized by the presence of multiple lesions.[10,18] Multiple cavernomas are found in approximately 10–20% of cases of cerebral cavernoma.[1,4,8,11,18] However, simultaneous recurrent bleedings in multiple lesions are rare. The risk of overt hemorrhage in cavernomas depends on several host and lesion factors: age, sex, location, associated venous malformation, and previous bleeding.[15] In addition, pregnancy and the puerperium may be associated with increased risk of bleeding and aggressive behavior in cavernomas.[15,19] Furthermore, environmental factors including irradiation and infection may interfere with the biological behavior of cavernomas.[5,16] Simultaneous hemorrhages from distinct lesions in this case may be predisposed by extralesional mechanisms such as hormone expression.

These brainstem cavernomas were considered to be ''kissing'' lesions since contact was observed on preoperative MR imaging including T_2-weighted imaging. However, a thin parenchymal layer between the lesions was observed intraoperatively. T_2-weighted imaging provides a false sense of proximity of the cavernoma because of the ferromagnetic properties of hemosiderin. The yellowish hemosiderin-stained gliotic parenchyma that surrounds the brainstem cavernoma should be considered as functioning tissue and should be preserved as far as possible.[6] Such a thin layer of functional parenchymal tissue may be compressed between ''kissing'' cavernomas in the brainstem. Therefore, to avoid unnecessary brainstem damage due to blind incision at the bottom, our first resection was limited to only the pontine tegmentum cavernoma in our case.

Several approaches for resection of pontine cavernoma have been used. Most cavernomas located at the pontine tegmentum, particularly with bulging to the fourth ventricle, have been resected through a trans-fourth ventricular floor.[2,6,12,20] Two safe entry zones to the brainstem were identified on the floor of the fourth ventricle: the supra- and infrafacial triangles.[9] However, this approach is not applicable if the cavernoma is localized ventrally in the pons. Resection of the ventral pontine cavernoma, especially if located laterally, is often performed through a lateral suboccipital retrosigmoid approach.[1,2,6,14] This approach has limited exposure of the anterior surface, so the combined retrosigmoid-subtemporal approach is also used.[1] Various microsurgical approaches for lesions in the anterior part of the pons have been reported, including the anterior petrosal, transcondylar fossa, far lateral, translabyrinthine, transcochlear, and transoral transclival approaches.[1,3,12,14,17,21] In this case, the trans-fourth ventricular floor approach for cavernoma in the pontine tegmentum is most suitable as the lesion is shallowest with this approach, but the choice of approach for ventral cavernoma is debatable. We selected the approach through the former removal route via the trans-fourth ventricular floor for the ventral pontine cavernoma for the following reasons: location in the lower pons, left corticospinal tract

compressed by the lesion laterally, adjacent to the previous removal cavity, and in the same approach direction as the resected lesion. This approach resulted in successful resection, particularly for resection of ''kissing'' lesions with difficult access in the brainstem, and is technically feasible with microsurgical procedures.

A few risks and limitations remain in this approach because of the previous removal route in the brainstem. In our patient, transient dysphagia was observed after the second operation, which was caused by damage to the left vagal nucleus during manipulation for resection, as illustrated by the enlarged entry on the floor of fourth ventricle on follow-up MR imaging. Care to prevent injury to the brainstem surrounding the entrance is important because this approach was a narrow corridor like a thin tube. Particularly in a trans-fourth ventricular floor approach, functionally important nuclei and tracts surround both triangles. Moreover, the removal cavity of the lesion may gradually shrink after surgery and the displaced functional structures may return to the former location. To avoid damaging the surrounding neuronal structures, this approach can be used in the same operation or in the early period following the first operation. In this case, second resection was scheduled soon after the residual cavernoma was identified on postoperative MR imaging, but the lesions could have been resected in one procedure using this approach if neuronavigation had been available at the first operation. In addition, if the positional arrangement of ''kissing'' lesions on MR imaging is not oriented on the same direction with the approach, only one of the ''kissing'' lesions can be resected, and a different entry point or approach to the brainstem should be selected.

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Address reprint requests to: Takahiro Murata, MD, Department of Neurosurgery, Shinshu University School of Medicine, 3–1–1 Asahi, Matsumoto 390–8621, Japan.