Technique

Surgery for mesencephalic cavernoma: case report

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Abstract

Background: Intrinsic midbrain tumor has been one of the most challenging therapeutic tasks in neurosurgery because of its prognosis and risks associated with surgical procedures. We encountered a rare case of cavernoma located in the anterior-mesencephalic region presented as parkinsonism tremor.

Case Description: A 42-year-old woman had a resting tremor for 10 years. This tremor involved the left shoulder and the proximal and distal limb, which was exacerbated by any attempted movement and became grossly uncontrollable. She showed a partial right ophthalmoplegia with mydriasis. Her right upper and lower extremities had normal strength, but her left extremities had three-fifth strength. Her sensation was intact. Magnetic resonance imaging demonstrated a popcorn-like rounded lesion in the right ventral midbrain adjacent to the medial cerebral peduncle. To access this anterior-medial portion of the midbrain, we chose a frontotemporal transsylvian route via an orbitozygomatic craniotomy. With a longitudinal pial incision between the frontopontine fibers and the pyramidal tracts in the peduncle, the cavernoma was totally removed en bloc. After the operation, the tremor dramatically disappeared. The muscle strength of her left lower extremity improved to four fifths, whereas the upper extremity was still the same. The preoperative left oculomotor palsy seemed to have no improvement.

Conclusions: Clinically manifested cavernoma due to repeated hemorrhage needs surgical intervention. With an optimal surgical approach, fairly safe entry zones on the anterior face of the rostral brainstem may be accessible, which provides a successful resection of a mesencephalic cavernoma without postoperative complications.

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Keywords: Cavernoma; Cerebral peduncle; Corticospinal fibers; Mesencephalon; Oculomotor nerve palsy; Surgical approach; Tremor

1. Introduction

Cavernomas have recently received a great deal of attention because of the improvement of neuroimaging and heightened clinical awareness. It accounts 5% to 10% of vascular malformations in the central nervous system. It is most commonly found in the cerebral hemispheres and rarely in the brainstem region with very low percentage in the mesencephalic region [6,13]. The surgical treatment of brainstem cavernomas is associated with an acceptable rate of postoperative morbidity compared with the higher risks of neurologic deficits that have been demonstrated after multiple hemorrhages in this delicate region. Nevertheless, decision making regarding the treatment of patients with mesencephalic cavernomas remains controversial because of the risks associated with surgery-related morbidity compared with the natural history of the disease.

To date, very few studies on the cavernoma located in the intrinsic midbrain have been focused on by the authors, though several authors have reported on the surgical treatment of brainstem cavernomas before [2-4,6,8,9,
20,22]. In the present article, a patient harboring an anterior midbrain cavernoma manifested by parkinsonism was reported. We discussed identifications of safe entry zones on the surface of cerebral peduncle as well as surgical techniques.

2. Case report

2.1. Clinical presentation

This 42-year-old Chinese woman had suffered left upper extremity tremor for 10 years. She had been treated as Parkinson’s disease with medication of L-dopa, however, the symptom was deteriorated continually. By the time of admission, she could not use her left arm because of this disabling tremor. She began to have a severe headache with diplopia 2 weeks before she was referred to our department. On examination, the patient had a resting tremor involving the left shoulder and the proximal and distal limb. On attempting posture, this irregular low-frequency tremor became grossly uncontrollable and was further exacerbated by any attempted movement. She showed a partial right oculomotor palsy presenting mild ptosis, limitations of medial and upward gaze, and dilated pupil with a negative light reflex. Her right upper and lower extremities had normal strength, but her left upper and lower extremities had three-fifth strength. Her sensation was intact to light touch throughout.

2.2. Radiologic findings

Magnetic resonance imaging demonstrated a popcorn-like rounded lesion on both T1- and T2-weighted images (Fig. 1). A rim of decreased signal intensity at the periphery of a heterogeneous central signal was characteristic on T2-weighted images.

2.3. The operative procedure

With brainstem-evoked potentials monitoring, the patient was placed supine. Her head was fixed in a 3-point Mayfield head frame with slight extension and 30° rotation to the contralateral side of the approach. An arc incision was made, which began 1 cm anterior to the tragus at the root of the zygomatic arch, curved forward and crossed to the contralateral mid-pupillary line.

The craniotomy was performed in the usual fashion with a high-speed pneumatic drill. A series of osteotomies were...
completed to free the lateral orbital roof and rim, the top of the malar eminence, and the zygomatic arch from the cranial base in a single piece (Fig. 2).

A semilunar dural opening was created. The sylvian fissure was then opened microsurgically. As the superficial sylvian and opticocarotid cistern were opened, cerebrospinal fluid was removed to promote brain relaxation and minimize the need for brain retraction. Arachnoid adhesions to the third and fourth cranial nerves and the edge of the tentorium were transected as the temporal lobe was retracted gently. By then, the midbrain and PCA were exposed in the center of the surgical field. With the operative microscope, the optical nerve, ICA, and MCA were visualized medially; the oculomotor nerve, PCoA, and BA anteriorly; and the SCA and trochlear nerve posteriorly (Fig. 3). After the PCA was then dissected away from the peduncle, a 1-cm longitudinal pial incision along the peduncle superolaterally to the oculomotor nerve was made. This incision was chosen at the conjunction between the anterior one third and the posterior two third of the cerebral peduncle. After dissecting 3 mm deeply, a thin layer of hemosiderin-stained neural tissue was encountered overlying the cavernous. When a boundary between the nidus and the parenchyma was well distinguished, the cavernoma was totally removed en bloc (Fig. 4).

Finally, the dura was closed and the orbitozygomatic flap was returned and secured to the skull (Fig. 5). The temporalis muscle and fascia were closed in separate layers. The scalp was closed in multiple layers.

2.4. Outcome

Histologic examination revealed multiple dilated cavernous spaces lined by endothelium with no intervening

![Fig. 3. The operative microscopy showing the midbrain and PCA in the center of the surgical field; the optical nerve (II), ICA, and MCA medially; the oculomotor nerve (III), PCoA, and BA anteriorly; the SCA and the tentorium edge laterally; and the trochlear nerve (IV) posteriorly. The pial incision on the surface of the ventral cerebral peduncle (indicated as a bar in the pictures) was chosen longitudinally between the CST and the FPT after the PCA was moved away from the peduncle.](image)

![Fig. 4. The cavernoma was being resected after identification of a hemosiderin-stained rim between the nidus and the surrounding parenchyma.](image)

![Fig. 5. The orbitozygomatic flap was returned and secured to the skull.](image)

![Fig. 6. Postoperative MRI delineating a complete excision of the mesencephalic cavernoma.](image)
brain tissue. The diagnosis of cavernoma was thereafter confirmed pathologically. Postoperatively, the tremor symptom was entirely solved. The muscle strength of her left lower limb improved to four fifths, whereas the upper limb remained the same. The preoperative left oculomotor palsy seemed to have no improvement. The postoperative MRI showed a completed excision of the mesencephalic cavernoma (Fig. 6).

3. Discussion

Cavernomas are vascular anomalies with cavernous spaces lined by endothelial cells and collagen, lacking smooth muscle or intervening neural tissue, filled with blood at various stages of degradation and surrounded by gliotic brain. The ventral midbrain is a rare location for cavernomas [5,11,21]. According the literature, brainstem cavernomas account for fewer than 20% of intracerebral cavernomas and are mostly found in the pons [13]. Samii et al [17] in 2001 reported 36 cases of brainstem cavernomas. Four of the cases were located in the posterior mesencephalic region, and none was observed in the anterior midbrain.

In the present case, according to the iconological delineation as well as the ophthalmoplegia and parapyramidal symptoms, we localized the nidus at the anterior portion of the substitute nigra, medially to the pyramidal tracts, laterally to the root filaments of oculomotor nerve, and anteriorly to the red nucleus (Fig. 7). In the operation, the cavernoma was found to have bled, though we did not know how many times it had happened during the past 10 years. A chronic compression and repeated hemorrhage in this precarious region contributed to the symptoms of the patient. Parkinsonism is an uncommon symptom of cavernomas. Only 1 case of cavernoma with tremor has been reported in the literature so far, which was a 56-year-old man with a cavernoma located in basal ganglion, who subsequently developed parkinsonism. The patient refused the surgical intervention and received L-dopa trial; however, no change in the tremor and bradykinesia was observed in spite of high doses of L-dopa [7]. Remy et al [16] reported 6 patients who harbored peduncular lesions (other than cavernomas) and manifested contralateral tremors. In the study, a marked decrease of the uptake of fluorodopa labeled with fluorine 18 in the striatum ipsilateral to the lesion was observed.

It is a challenge to treat midbrain cavernomas. Radio-surgery has been used in the treatment of cavernomas in the delicate region of brainstem; however, the incidence of radiation-induced complications is considerably higher [10]. In virtue of the surrounding gliotic layer of the cavernoma, excision of the nidus without damage to the neural structures is theoretically feasible. The benefit of surgery on the treatment of brainstem cavernomas has already been reported [1,6]. Numerous authors have stressed that the complete removal of brainstem cavernoma in patients with recurrent symptomatic hemorrhages is the only choice to avoid further devastating rebleeding-related neurologic impairment [3,12,14,15,17,18,21]. Samii et al [17] believed that the incidence of new postoperative cranial nerve deficits is clearly lower than that demonstrated preoperatively because of the brainstem hemmorhages.

With an appropriate surgical approach and sophisticated dissection, satisfactory results could be obtained. During the resection, protection of the CST and preservation of the motor function are the neurosurgeon’s primary concerns. In the present case, the shortest way to the nidus is from ventral mesencephalon. Anatomically, the root filaments of oculomotor nerve, the frontopontine fibers, the corticospinal fibers, and the parietotemporal fibers rank medial-lateral in the cerebral peduncle. To save those axonal bundles as much as possible, we made a longitudinal pial incision between the frontopontine fibers and the pyramidal tracts in the peduncle. To access this medial target, we chose a frontotemporal transsylvian route via an orbitozygomatic craniotomy. Increasing the amount of bone removed by using an orbitozygomatic approach instead of a pterional approach converts a narrow space into a wide portal, allowing surgeons to work closer to the cerebral peduncle while decreasing the need for brain retraction. According to the cadaver study by Schwartz et al [19], the areas of exposure for the posterior clinoid, edge of tentorium, and basilar tip targets were increased 26% to 39% with orbital rim osteotomy and an additional 13% to 22% with removal of the zygomatic arch.

The timing of surgery posthemorrhage has been also a matter of debate. Fahlbusch et al [9] have advocated resection of the lesion in the subacute phase after stabilization of a patient’s neurologic condition to avoid reactive gliosis, which may occur months after extralesional bleeding. Nevertheless, the study of Samii et al [17] demonstrated that there was no statistical difference in

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Fig. 7. Sketch showing the anatomical transect of the midbrain and the surgical approach. The FPT, CST, and PTT ranked on the sequence from anterior-medial to posterior-lateral in the cerebral peduncle. The cavernoma is located at the medial-anterior midbrain between the CST and FPT and in front of SN. An incision between the CST and FPT was created on the surface of cerebral peduncle.
favor of patients who underwent surgery within 3 months posthemorrhage compared with those in whom resection was performed after 3 months.

4. Conclusion

A mesencephalic cavernoma mainly presented with parkinsonism was a very rare case. Those clinically manifested cavernomas need surgical resection. An optimal surgical approach should be selected as the surrounding vital structures are not compromised. To obtain a good operative outcome for the cavernoma at the ventromedial part of the brainstem, we believe the following points are very important: (a) A precise localization of the lesion with its anatomical relationship to pyramidal and parapyramidal tracts should be defined preoperatively based on the clinical presentation and neuroimaging delineation. (b) A frontotemporal transsylvian route via an orbitozygomatic craniotomy is an appropriate surgical approach to access the cerebral peduncle region, which provides better operative trajectory, shallower surgical field, and less brain retraction. (c) To save the CST, a longitudinal pial incision between the frontopontine fibers and the pyramidal tracts in the peduncle is recommended. (d) A hemosiderin-stained boundary between the nidus and the brain parenchyma should be identified before removing the cavernoma, which offers advantageous dissection and total resection of the lesion without damage to the surrounding vital structures. (e) It is always wise to resect a cavernoma en bloc with minimal coagulation.

References


Commentary

The authors on this article are to be complimented for the excellent result they achieved operating on this difficult case. Through a frontal temporal transsylvian route via an orbitozygomatic craniotomy, they obtained a good exposure of the ventral lateral right mesencephalon and entered the peduncle between the frontal pontine fibers and the pyramidal tracts, safely removing the cavernoma without adding any neurologic deficits. In fact, whenever the ventral midbrain needs to be entered, as we highlighted in 1995 [1], we must take into account that the most important anatomical landmark of this area is the emergence of the third cranial nerve with the PCA above. Below the nerve, the SCA runs medial to lateral, and the safe entry zone into the midbrain is the small rectangular area outlined medially by the exit of the third cranial nerve and the BA, inferiorly by the SCA, superiorly by the PCA, and laterally by the tentorial edge. This narrow but fairly safe window allows surgical access through the more medial part of the peduncle, sparing the motor tract, which occupies only the intermediate three fifth or so of the peduncle. In this case, the surgeons used the safest entry zone, and this explains the success of the operation. Without taking anything away from what the authors say in this contribution, I would like to add that the tremor mentioned in more than 1 place, defined as parkinsonism, is more likely to be a Holmes