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Case Report

A rare case of rebleeding brainstem cavernoma in a 5-month-old-girl

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ABSTRACT

Background: Brainstem cavernomas (BSCs) are relatively rare intracranial vascular lesions that, if left untreated, can be devastating to the patient. The lesions are associated with a myriad of symptoms, depending on their size and location. However, medullary lesions present acutely with cardiorespiratory dysfunction. We present the case of a 5-month-old child with a BSC.

Case Description: A 5-month-old child presented for the 2nd time with sudden respiratory distress and excessive salivation. On the first presentation, brain magnetic resonance imaging (MRI) showed a 13 × 12 × 14 mm cavernoma at the pontomedullary junction. She was managed conservatively but presented 3 months later with tetraparesis, bulbar palsy, and severe respiratory distress. A repeat MRI showed enlargement of the cavernoma to 27 × 28 × 26 mm with hemorrhage in different stages. After hemodynamic stabilization, complete cavernoma resection was performed through the telovelar approach with neuromonitoring. Postoperatively, the child recovered motor function, but the bulbar syndrome persisted with hypersalivation. She was discharged on day 55 with a tracheostomy.

Conclusion: BSCs are rare lesions that are associated with severe neurological deficits due to the compactness of important cranial nerve nuclei and other tracts in the brainstem. Early surgical excision and hematoma evacuation for superficially presenting lesions can be lifesaving. However, the risk of postoperative neurological deficits is still a major concern in these patients.

Keywords: Brainstem hemorrhage, Brainstem, Bulbar palsy, Cavernoma, Telovelar approach

INTRODUCTION

Cavernous malformations, also known as cavernous angiomas, cavernous hemangiomas, or cavernomas, are well-circumscribed, low-flow, and angiographically occult lesions with no large arterial inflow or draining veins.[4] Brainstem cavernomas (BSCs) are rare, and their natural history and pathology are still unclear. Bleeding or rebleeding of BSC often causes severe neurological deficits, and their location and characteristics make surgical treatment a

challenge.[6] BSCs account for 18–35% of central nervous system cavernomas, with 57% occurring in the pons, followed by the midbrain (14%), pontomedullary junction (12%), and medulla (5%).[5] The telovelar approach and its modifications are widely used to remove lesions of the fourth ventricle and brainstem.[1] Total excision of BSCs remains the primary goal of microsurgery, as the risk of rebleeding following incomplete removal is significant. The removal of BSC is carried out by minimal brain incision and the shortest approach using functional radiological study, intraoperative navigation systems, and motor functional mapping to guide the trajectory of the approach.[4] We present a case of 5-month-old child with a BSC who presented with severe medullary dysfunction.

CASE DESCRIPTION

A 5-month-old girl was admitted to the pediatric intensive care unit (ICU) with dyspnea, stridor, cyanosis, pronounced weakness, lethargy, and a progressively decreasing level of consciousness until she was comatose. This was the second admission to the ICU.

First admission

At 3 months old, the patient was admitted to the ICU on mechanical ventilation following sudden respiratory distress and excessive salivation. A head and neck computed tomography scan and magnetic resonance imaging (MRI) [Figure 1] on admission revealed a brainstem hemorrhage and a giant suprascapular and mediastinal lymphangiomatous malformation. A second MRI with contrast (18 days later) revealed a cavernous malformation with a 13 × 12 × 14 mm hematoma in the right pontomedullary region with mild ventriculomegaly. The child was managed conservatively for 1 month and discharged after stabilization and resolution of her neurological deficits.

During the second admission, a brain MRI on admission revealed an increase in the size of the brainstem hematoma to 27 × 28 × 26 mm compared to the previous admission [Figure 2].

On examination, the patient was in stupor with a four score of 10, with tetraparesis, bulbar palsy, and severe respiratory distress. In addition, she had metabolic and electrolyte disorders and anemia (Hb 80 g/L) with thrombocytosis (641 × 10*9/L).

Surgical technique

With the patient in the prone position, and the head fixed in a Mayfield 3 pin at maximum neck flexion, a standard telovelar approach to the brainstem through a midline suboccipital craniotomy was performed. The dura was opened in a curvilinear form, followed by dissection of the arachnoid between the tonsils to expose the telovelar membrane. With gentle tonsillar retraction, the telovelar membrane was opened, and the floor of the fourth ventricle was exposed. The dorsal surface of the upper medulla oblongata was seen bulging and hemosiderin-stained [Figure 3a]. Brainstem mapping was performed using a monopolar probe.

A small incision was made at the most presenting surface of the cavernoma, and the hematoma was evacuated and sent for histology [Figure 3b]. The rest of the cavernoma was carefully resected together with the adherent capsule, as guided by neuromonitoring. The cavernoma was also sent for histopathological examination.

There was no significant reduction in the brainstem-evoked potentials, and there was minimal iatrogenic injury to the neural tissue. After complete excision of the cavernoma, hemostasis was achieved using thin Surgicel slices, duroplasty was performed using NeoDura and Hemopatch grafts, and closure was performed as standard.

In the early postoperative period, the patient's neurological functioning gradually improved. The patient was gradually weaned off of mechanical ventilation and transferred to auxiliary lung ventilation. By day 5, the patient had hemodynamically stabilized with a four score of 13. However, extubation was delayed considering the excessive salivation of approximately 500 mL/day, persistent bulbar palsy, and the patient's age. A tracheostomy was performed on day 22 to manage the bulbar palsy. The child was discharged

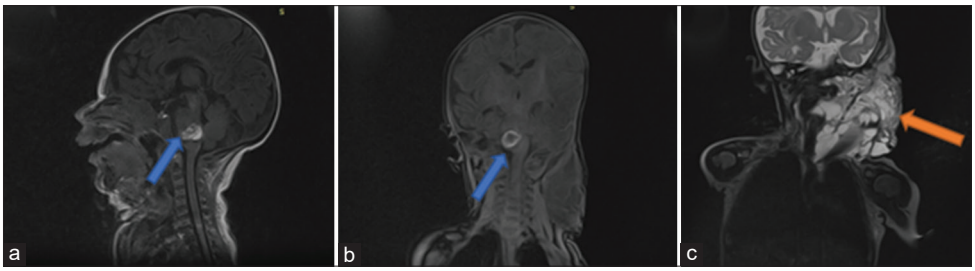

Figure 1: The magnetic resonance imaging on the first admission. The classic popcorn cavernoma (blue arrow) is seen in the right pontomedullary region of the brainstem in the sagittal (a) and coronal views (b). (c) The lymphangioma is seen in the left supraclavicular and neck region (orange arrow).

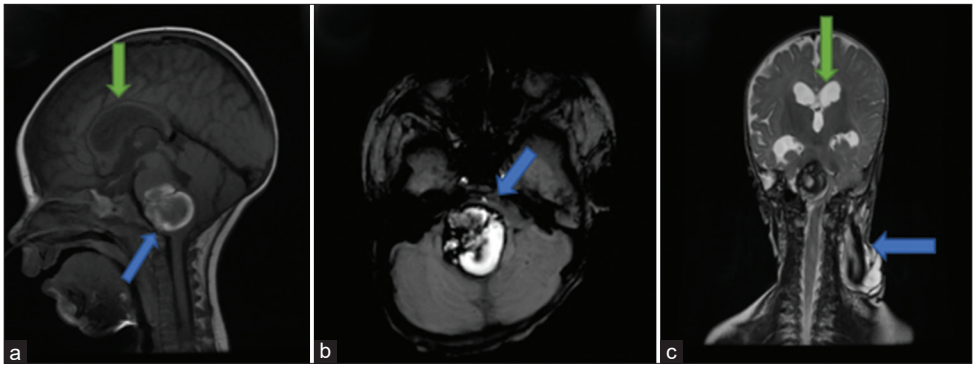

Figure 2: Brain magnetic resonance imaging on the second admission. (a) Sagittal cut showing a 27 × 28 × 26 mm pontomedullary cavernoma (blue arrow) and hydrocephalus (green arrow). (b) Axial cut showing the heterogeneous lesion with a hemosiderin ring and severe compression of the brain stem (blue arrow). (c) Coronal cut showing hydrocephalus (green arrow) and significant regression of the lymphangioma (blue arrow) compared to magnetic resonance imaging on first admission.

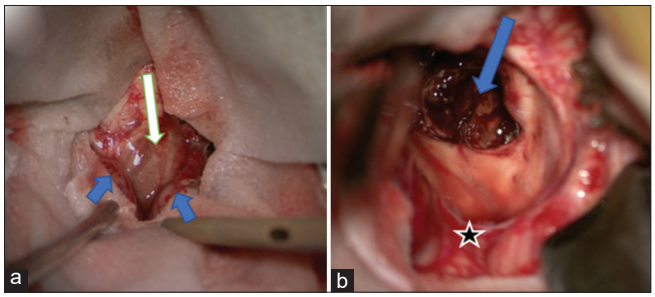

Figure 3: Intraoperative images. (a) The exposure of the telovelor membrane and the bulging medulla oblongata with hemosiderin discoloration (white arrow). The cerebellar tonsils (blue arrows). (b) The opened cavernoma with the preserved developmental venous anomaly (DVA) (blue arrow). Cerebellar vermis (black star).

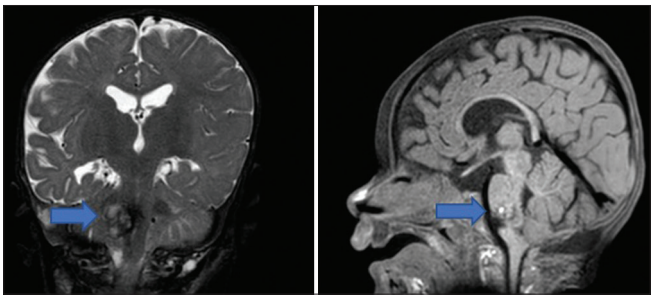

Figure 4: The 4 months postoperative magnetic resonance imaging. Blue arrow-resolving postoperative changes in the brainstem.

on hospital day 55 to continue outpatient treatment and rehabilitation. The 4 months postoperative MRI showed normal resolving postoperative changes [Figure 4].

Histopathology showed the typical hamartomatous cystic dilation of the single layered vascular spaces. Molecular and genetic studies showed no mutations associated with familial cavernomas like Krit 1.

DISCUSSION

Cavernous malformations are the most prevalent type of vascular malformation in the brain, with an incidence of 0.4–0.5% and a prevalence of 0.6/100,000 inhabitants. The condition is more prevalent in males between the ages of 20 and 40 and is found in approximately 25% of children. The dynamic nature of cavernous malformations, including enlargement, regression, and *de novo* formation, can be attributed to different stages of hemorrhage and hemorrhage resolution, which can be seen in characteristic MR images.[15] Although it is estimated that around 20% of cavernous malformations occur in the brainstem, some studies have reported that this figure may be as high as 35%.[16]

The clinical course of brainstem cavernous malformations in children is highly variable, ranging from benign lesions to highly aggressive lesions with repetitive hemorrhages. Patients with brainstem cavernous malformations may present with cranial nerve deficits, sensory changes, headache, motor deficits of the extremities, diplopia, ataxia, and vertigo. Cranial nerve deficits are the most common early clinical presentation.[1]

Assessing symptoms in infants can be challenging, given the severity of the disease and the resulting neurological deficits.[14] In the case presented in this study, the patient exhibited hypersalivation, bulbar palsy, and tetraparesis, which are typical of a lesion in the lower brainstem. As discussed in the literature, despite the location of the lesion, there is usually a significant return of neurological function, and the tetraparesis and bulbar syndrome have resolved significantly.

Rebleeding is more frequent in conservatively treated patients or those with incomplete cavernoma resection. The rebleeding rate is 10%/year, which is considered

high in comparison with other cerebral vascular malformations.[2,8,11,12] Some studies have reported that the size of the lesion (>20 mm) is an independent risk factor for rebleeding.[2,9,10] Most recurrences occur within the first 5 years after the first bleed, with the highest rebleeding rates being reported in the first 2 years, followed by a gradual reduction in the risk of rebleeding over time.[2]

Given the longer life expectancy and greater functional recovery in children, surgical treatment should be considered early in young patients presenting with surgically accessible lesions and an aggressive clinical course.[13] Microsurgical treatment of BSCs immediately eliminates the risk of re-hemorrhage.[6] Other indications for surgical management of BSCs include: (1) progressive neurological impairments; (2) the clear presence of sudden or gradual bleeding on MRI, either within or outside cavernous malformations, resulting in pressure on surrounding tissues; and (3) the presence of a cavernoma/hematoma in close proximity to the brainstem surface, with <2 mm of brain tissue separating the cavernoma/hematoma and the outermost layer of the brain (pial surface). Severe clinical manifestations such as coma, respiratory issues, or unstable cardiac conditions do not preclude surgical intervention.[14]

The telovelar approach is a less traumatic surgical approach that uses natural planes. Through a C1 laminectomy, the rostral extent of the intraventricular exposure can be expanded, giving more flexible inferior-to-superior working angles. Moreover, the telovelar approach offers a corridor through noneloquent arachnoid planes and a safe and adequate working environment.[4,5,7] Unless otherwise indicated, no resection of the cerebellum is required with this approach.

Using this approach with functional radiological studies, intraoperative navigation, and motor functional mapping allowed for precise resection of the lesion with minimal damage to the surrounding neural tissue. In addition to neuromonitoring, there are safe entry zones to the brainstem that has been shown to be safe and effective in avoiding injury to the vital nuclei and tracts.[3] Analysis of these anatomical approaches is beyond the scope of the case report.

The patient regained significant motor function but had persistent bulbar syndrome on discharge, which was expected considering the size and location of the lesion. Close follow-up after surgery is essential for the early detection of potential complications, such as rebleeding, neurological deficits, or recurrence of the lesion. Imaging studies, such as MRI, can help monitor the progression of the lesion and detect any changes that may require further intervention.

CONCLUSION

BSCs are rare lesions that are associated with severe neurological deficits due to the compactness of important cranial nerve nuclei and other tracts in the brainstem. Early surgical excision and hematoma evacuation for superficially presenting lesions can be lifesaving. However, the risk of postoperative neurological deficits is still a major concern in these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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