

Atypical Hemorrhagic Presentation of a Fourth Ventricle Subependymoma: Case Report

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Abstract

To present a case of a fourth ventricle subependymoma (SE) with a spontaneous acute subarachnoid intra-cisternal bleeding. A 33-year-old man was admitted with 5 days history of oppressive occipital headache and neck pain without additional neurological focus. Unenhanced computed tomography (CT) scan demonstrated an isointense mass located in the fourth ventricle with a spontaneously hyperdense acute extratumoral hemorrhage in the cisterna magna. Contrast-enhanced magnetic resonance imaging (MRI) revealed a well-delimited non-enhanced tumor, hypointense on T₁-weighted and hyperintense on T₂-weighted images, involving the floor of the fourth ventricle and extending caudally into the cervical spinal canal via foramen magnum. Intraoperative, a large blood clot was removed and a macroscopically hypovascular lesion was completely excised from the right lateral recess and the floor of the fourth ventricle. Intra and postoperative immuno-histopathological examination revealed a SE. The patient has a normal postoperative course and was discharged in the fifth postoperative day. A 10-month postoperative MRI study confirmed a complete tumor resection. Symptomatic SEs should be surgically treated emphasizing the urgency in the presence of hemorrhage. The interest of this case is to demonstrate that infratentorial SEs although extremely rare, might present with acute subarachnoid bleeding.

Key words: cisterna magna hemorrhage, fourth ventricle subependymoma, hemorrhagic subependymoma

Introduction

Subependymomas (SEs) are ependymal tumors developed from subependymal glial precursors and characterized by an asymptomatic benign clinical course and distinguished from the ordinary ependymomas by their general better prognosis. These rare tumors are more frequent in men predominantly adults with a general incidence accounting from 0.2% to 0.7% of intracranial tumors.^{8,10,11,14,15} They commonly arise along the ventricular system walls, encountered in the 50–60% of the cases in the fourth ventricle, followed by the lateral ventricle 30–40%, and less frequently, in the septum pellucidum area and spinal cord.^{13,16} If symptoms are present, there are generally related with the obstruction of cerebrospinal fluid (CSF) pathway.^{6,8,9} Tumoral-related hemorrhage represents an extremely rare presentation sign generally associated with intralesional bleeding that may cause acute and severe neurological deficits.² We present a case of a fourth ventricle SE with a spontaneous acute extra tumoral subarachnoid

intra-cisternal bleeding. We perform a literature review of this unusual presentation.

Case Report

A 33-year-old man, with no relevant clinical record, was admitted in our institution with 5 days history of oppressive occipital headache and posterior cervical pain. Neurologic examination revealed photophobia and neck stiffness without additional neurological focus. Laboratory test showed normal coagulation parameters. Unenhanced computed tomography (CT) scan demonstrate a well-circumscribed isointense mass located in the inferior portion of the fourth ventricle with a spontaneously hyperdense acute extratumoral hemorrhage in the cistern magna (Fig. 1). No vascular abnormalities were revealed on CT angiography. Contrast-enhanced magnetic resonance imaging (MRI) demonstrated a well-delimited non-enhanced tumor, hypointense T₁-weighted and hyperintense on T₂-weighted images, involving the floor of the fourth ventricle extending caudally into cervical spinal canal via foramen magnum measuring approximately 30 mm in diameter (Fig. 1).

The patient was operated immediately through a midline suboccipital craniectomy and a velotonsillar approach. The cistern magna was opened and hemorrhagic CSF under high pressure was aspirated sequentially. A large blood clot was removed and a firm, well delimited, yellow-white

partially cyst-lobulated, macroscopically hypovascular lesion was recognized and completely excised, which was attached to the right lateral recess and the floor of the fourth ventricle previous desvascularization from small branches of the right posteroinferior cerebellar artery (Fig. 2). Intra and postoperative immunohistopathological examination revealed a SE (Fig. 3). The patient has a normal postoperative course and was discharged on the fifth postoperative day. A 10-month postoperative MRI study confirmed a complete resection of the tumor with normal CSF circulation (Fig. 4).

Discussion

These benign indolent growth lesions are commonly smaller than 2 cm in diameter, larger tumors tend to cause symptoms related to their location.^{8,14} Clinical manifestations were relatively more often caused by supratentorial lesions, 66% of these cases were symptomatic compared to 36% of patients with tumors of the fourth ventricle.¹⁶ Large series of fourth ventricle SEs reports clinical manifestation only in one-third of the patients, especially in those arising from the floor of the fourth ventricle.¹⁶ Loss of balance, vertigo, headache, and vomiting were the most common presenting symptom usually caused by CSF obstruction.^{7,14,15} Hemorrhage is an extremely rare sign in SEs. To our knowledge only 12 cases have been reported till date (Table 1). The etiology remains uncertain, only 4 patients had predisposing factors such as arterial hypertension, vascularized tumor, and anticoagulant therapy.^{2,4,9,18}

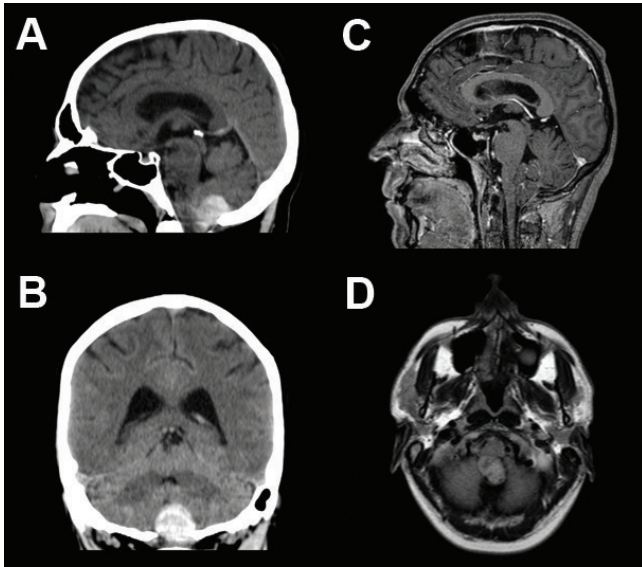


Fig. 1 A, B: shows sagittal and coronal CT views demonstrating acute extratumoral hemorrhage between the cisterna magna and the atlas. C, D: represents sagittal and axial views of a non-enhancing fourth ventricle tumor on T₁-weighted contrast MRI sequences. CT: computed tomography, MRI: magnetic resonance imaging.

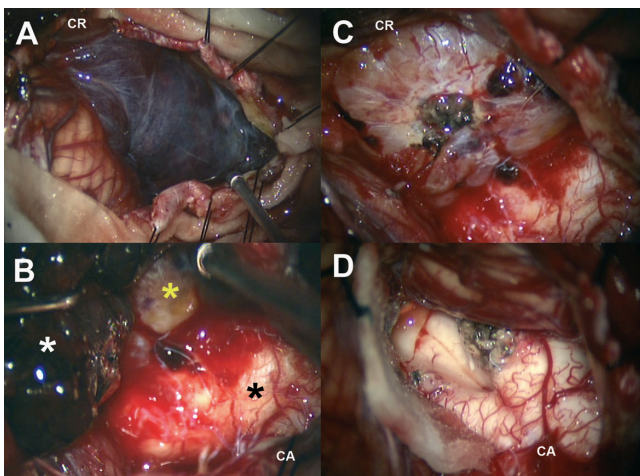


Fig. 2 A: intraoperative photo demonstrate subarachnoid hemorrhage (CR, cranial). B: white asterisk shows large blood clot, yellow revealed fourth ventricle tumor and the black asterisk mark the brain stem (CA, caudal). C: represents tumor site of hemorrhage. D: shows complete tumor excision and the fourth ventricle floor.

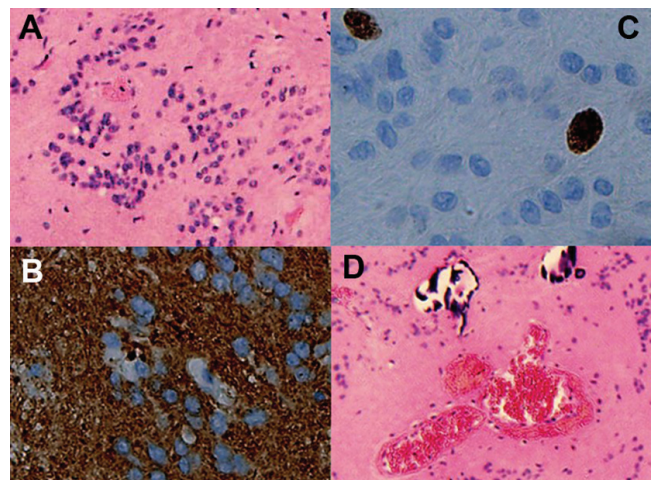


Fig. 3 A: represents clusters of isomorphic nuclei embedded in a dense fibrillary matrix of glial processes and an ependymal pseudorosette (H&E 10 \times). B: shows immunoreactivity for GFAP (40 \times). C: revealed Ki67 label index below 2% (40 \times). D: demonstrates some lobular architecture, in a hypocellular area in a highly fibrillary background area with calcifications (H&E 10 \times). H&E: hematoxylin-eosin, GFAP: glial fibrillary acidic protein.

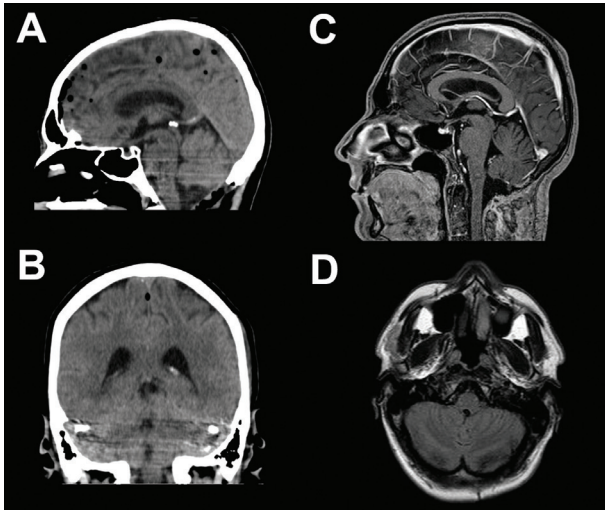


Fig. 4 A, B: shows sagittal and coronal postoperative CT views demonstrating surgical approach and hemorrhage resolution. C, D: represents T₁-weighted contrast MRI sagittal and axial views demonstrating complete tumoral resection. CT: computed tomography, MRI: magnetic resonance imaging.

although some authors postulate the hypothesis that the acute extratumoral bleeding may be the result of tearing surrounding veins during tumoral growth.^{17,21)}

Only 4 cases with subarachnoid hemorrhage (SAH) have been reported in all of them in supratentorial location.^{3,4,9,12)} Our case presents SAH and a large blood clot in the cisterna magna without any evidence of local veins injury but a hemorrhagic focus on tumor surface suggesting the possibility of bleeding from a tumor vessel spontaneously or as result of a traumatic mechanism due to the fact that the lower part of tumor is below the foramen magnum (Figs. 1, 2). To our knowledge, this is the first case of spontaneous acute hemorrhage in an infratentorial SE reported in the literature.

SEs first differential diagnosis are ordinary ependymomas, the former tend to extend beyond the ventricular margins, enhance markedly, and demonstrate more frequently degenerative changes such as cyst formation and calcifications.^{5,10,14,15)} The pathognomonic diagnosis is due to histopathological examination where SEs shows features of astrocytic and

Table 1 Case reference of hemorrhagic presentation of subependymomas

| Case no. | Author/Year | Age/Gender | Clinical presentation | Predisposing factor | Location/Size (cm) | Hemorrhage | Surgical approach/Resection | Complication/Outcome |
|----------|---|------------|----------------------------|----------------------------|--------------------|--------------|-----------------------------|--------------------------------------|
| 1 | Scheithauer (1978) ¹⁶⁾ | 81/F | HA/AC | NA | LV/ Large | IT | NOP | Massive hemorrhage/died |
| 2 | Changaris et al. (1981) ³⁾ | 16/M | HA/Blurred vision | No | AT,OH/7 | IT, SAH | POTC/Total | Homonymous hemianopsia/Good |
| 3 | Seiki et al. (1984) ¹⁷⁾ | 33/F | HA/AC | No | AT/ NA | IVH | POTC/Partial | NA |
| 4 | Yamasaki et al. (1989) ²⁰⁾ | 54/F | HA | No | FH/5 | IT | TC/Total | Transient memory impairment/Good |
| 5 | Marra et al. (1991) ¹²⁾ | 42/F | HA | NA | FH/2.5 | IVH, SAH | TC/Total | No/Good |
| 6 | DiLorenzo et al. (1991) ⁴⁾ | 46/M | HA | AHT | FH/4 | IVH, SAH | FTC/Total | No/Good |
| 7 | Lindboe et al. (1992) ⁹⁾ | 63/M | Disorientation memory loss | High vascularized tumor | FH/5 | IT, IVH, SAH | TC/Partial | Re-bleeding /Died |
| 8 | Viale (1994) ¹⁹⁾ | 52/M | HA/AC | NA | FH/3 | IT | NA/Total | No/Good |
| 9 | Furie and Provenzale (1995) ⁵⁾ | 46/M | HA | NA | LV/2 | IT | NA/NA | NA |
| 10 | Carrasco et al. (2010) ²⁾ | 71/M | AC | AHT/ Anticoagulant therapy | FH/3 | IVH | F ^o TC/Total | Memory Impairment/Good |
| 11 | Akamatsu et al. (2010) ¹⁾ | 32/M | HA/AC | No | LV/2 | IVH | F ^o TC/total | No/Good |
| 12 | Sharma et al. (2010) ¹⁸⁾ | 25/M | HA/ Blurred vision | Vascularized tumor | LV, AT/ NA | IT | POTC/Total | Seizures, altered consciousness/Good |
| 13 | Present case | 32/M | HA/Posterior cervical pain | No | 4thV/3 | ICM, SAH | SOVT/total | No/Good |

AC: altered consciousness, AHT: arterial hypertension, AT: atrium, F: female, FH: frontal horn, 4th V: fourth ventricle, FTC: frontal transcallosal, HA: headache, ICM: intra cisterna magna, IT: intratumoral, IVH: intraventricular hemorrhage, LV: lateral ventricle, M: male, NA: not available, NOP: not operated, OH: occipital horn, POTC: parieto-occipital transcortical, SAH: subarachnoid hemorrhage, SOVT: suboccipitalvelo-tonsilar, TC: transcallosal.

ependymal differentiation with small clusters of nuclei and hypocellular areas in a highly fibrillary background, however mixed forms of SEs with ependymomas are rather common. Immunohistochemically, SEs stain diffusely with S-100 protein and glial fibrillary acidic protein (GFAP) as demonstrated in Fig. 3.

Intraoperatively SEs are generally described as hypovascular or avascular tumors, although highly vascularized cases have been reported.^{9,10} The presence of a tumor with SAH does not necessarily indicate a malignant tumor,³ but could represent a life-threatening condition with increased risk for pre- and postoperative hemorrhage and justifies a thorough surgical planning and a promptly intervention. Large SEs often have several sites of attachment, but on the other hand these tumors can be potentially cured by surgery, a complete excision must always be attempted avoiding the damage to adjacent vital structures.

Conclusion

SEs of the fourth ventricle are commonly benign asymptomatic slow growing lesions. Symptomatic tumors should be treated surgically emphasizing the urgency in the presence of hemorrhage. The interest of this case is to demonstrate that infratentorial SEs although extremely rare, might present with acute subarachnoid bleeding.

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