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Case Report

Spinal dural arteriovenous fistula rupture after Rathke's cleft cyst endoscopic resection: Case report and literature review

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ABSTRACT

Background: Spinal dural arteriovenous fistula (SDAVF) is the most frequent vascular malformation of the spine and accounts for approximately 70% of all vascular spinal malformations. In rare cases, SDAVF rupture and subsequent subarachnoid hemorrhage or intramedullary hematoma may occur. The aim of this article is to present a fatal case of SDAVF rupture after a Rathke's cleft cyst (RCC) endoscopic resection.

Case Description: An 80-year-old female was referred to our hospital with a clinical presentation of bilateral reduction in visual acuity, bitemporal hemianopsia, and sellar magnetic resonance imaging (MRI) highly suggestive of RCC. After the first endonasal endoscopic surgery, the cyst was partially removed and vision improved. No signs of cerebrospinal fluid (CSF) leak were observed. After 1 year, the patient returned because of RCC recurrence and decreased visual acuity. In the second procedure, the lesion was totally resected and CSF leak was observed. A nasoseptal flap was rotated to cover the skull base defect. The patient developed subtle paraparesis followed by paraplegia on the 4th postoperative day. The dorsal spine MRI revealed a T3-T4 intramedullary hematoma. A dorsal laminectomy was performed and a SDAVF was observed. During microsurgery, at the right T3 nerve root level, an arteriovenous shunting point was identified, coagulated, and divided. The intramedullary hematoma was evacuated. The patient developed neurogenic and septic shock and died.

Conclusion: Venous hypertension, venous wall fragility, and venous thrombosis seem to be the main factors involved in SDAVF rupture. In this particular case, reduction of the extravascular pressure and sudden variation in the pressure gradient caused by sustained CSF leak, also appeared to play an important role in SDAVF rupture. It may represent one more complication related to radical resection of RCC.

Keywords: Cerebrospinal fluid fistula, Rathke's cleft cyst, Rupture, Spinal dural arteriovenous fistula, Transsphenoidal resection

INTRODUCTION

A spinal dural arteriovenous fistula (SDAVF) is an abnormal connection between arteries and veins located near the dura mater.[15] They are the most frequent vascular malformation of the spine and account for approximately 70% of all vascular spinal malformations.[11] The SDAVF

tends to be located dorsally in the low thoracic and lumbar area with 80% of cases occurring between T6 and L2.[15] Arterialization of the medullary vein leads to venous congestion in the coronal venous plexus of the spinal cord. As venous pressure increases, there is decreased tissue perfusion along with vascular steal and ischemia. In rare cases, rupture of the SDAVF fistula and subsequent subarachnoid hemorrhage or intramedullary hematoma may occur.[18] The aim of this article is to present a fatal case of SDAVF rupture after an endoscopic Rathke's cleft cyst (RCC) radical resection. We did not find any report describing this association in medical literature analysis and retrieval system online basis as of June 2021.

CLINICAL PRESENTATION

An 80-year-old female was referred to our tertiary hospital for surgical treatment of a recurrent RCC. The first endoscopic surgery, performed 2 years previously in the same institution, consisted of cyst fenestration, and partial cyst wall resection. The pedicled nasoseptal flap technique[9] was used to reconstruct the skull base. No signs of cerebrospinal fluid (CSF) fistula were observed. At that time, the clinical presentation was bilateral reduction in visual acuity, bitemporal hemianopsia, and mild bifrontal headache for 1 year. There was no pituitary disfunction. Preoperative sellar computed tomography (CT) and magnetic resonance imaging (MRI) scan revealed a cystic sellar lesion [Figure 1a-c]. After resection, she recovered from visual disturbances and managed to return to her daily activities.

One year later, the patient presented recurrence of visual disturbance and bifrontal headaches. The neurologic examination revealed anisocoric pupils, with fixed mydriasis in the left eye and bitemporal hemianopsia. A new endoscopic transnasal transsphenoidal surgery was performed and a gross total resection (GTR) was achieved. However, during cyst dissection, a CSF leak was observed. Resurface of the skull base with a nasoseptal flap was performed. No hypertensive episodes were observed during the procedure. Postoperative sellar MRI confirmed total resection [Figure 2a-c]. However, there was a "reservoir sign" in which the CSF goes out when taking a head up position in the lying position. Due to the low CSF leakage, we chose a conservative approach with head elevation and neuro check. This condition remained stable until the 3rd postoperative day and due to the minimum CSF outflow, no surgical procedure was performed.

The patient developed subtle paraparesis followed by paraplegia on the 4th postoperative day. Dorsal spine MRI revealed a T3-T4 intramedullary lesion. The lesion was hyperintense on T2 and most notably in the right side of the spinal cord on the axial plane [Figure 3a and b]. An emergency thoracic laminectomy (T2-T5) was performed [Figure 4a-c]. After dural opening, an SDAVF and an intramedullary hematoma were observed. The feeder artery on the right T3 dural sleeve was identified. The arterial supply was interrupted by coagulation of the epidural arteries around the right T3 nerve root. The vessels draining the dural fistula entered the dura as a single arterialized vein, which drained into a huge dilated tortuous spinal vein on the dorsal surface of the spinal cord. The draining vein-like varix was almost thrombosed and was partially projected into the cord. The lesion was classified as an intradural dorsal fistula according to Spetzler *et al*. classification[19] or Type I spinal arteriovenous fistula according to Di Chiro and Wener classification.[3] The shunting point was clearly identified, coagulated, and divided. The intramedullary hematoma was evacuated. The hematomyelia was assumed to have been caused by the rupture of the dilated intraparenchymal draining vein.

Immediately after surgery, the patient presented neurogenic shock needing high doses of norepinephrine. She was admitted to the intensive care unit. From this moment, no signs of CSF rhinorrhea were observed. Four days after laminectomy, there was a progressive worsening of neurogenic shock associated with septic shock due to pneumonia requiring higher doses of vasoactive amines. Mean arterial pressure ranged between 50 and 60 mmHg despite high doses of norepinephrine. Ten days after laminectomy, the patient developed a hypoxic encephalopathy resulting in brain death. The brain CT demonstrated a diffuse white

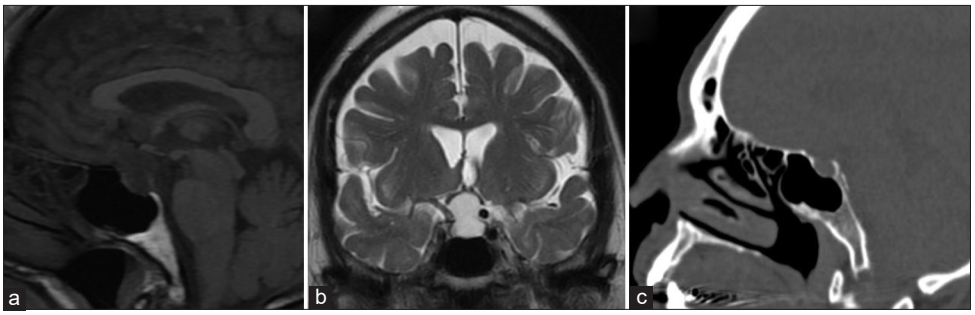

Figure 1: Preoperative sellar MRI and CT scan. (a) MRI - T1-weighted sagittal slice demonstrating hypointense cystic lesion in sellar space. (b) MRI - T2-weighted coronal slice demonstrating hyperintense lesion (c) CT sagittal slice demonstrating sellar type of sphenoid sinus.

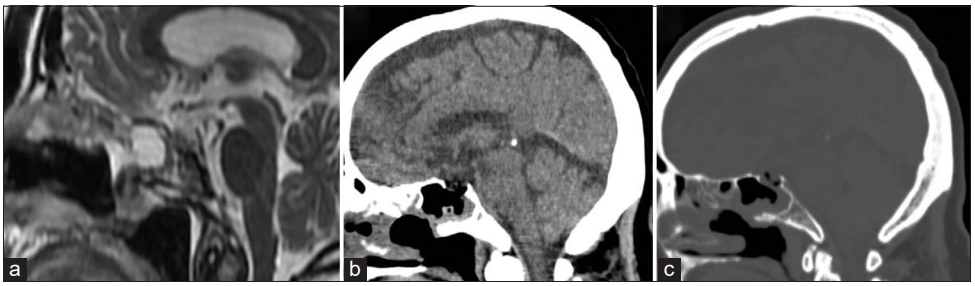

Figure 2: Postoperative sellar MRI and CT scan. (a) MRI - T2-weighted sagittal slice demonstrating empty sellar space after Rathke's cleft cyst gross total resection and nasoseptal flap closure. (b) CT sagittal slice shows minimum pneumoencephalon in sellar region after endoscopic procedure (c) Bone window CT sagittal slice - demonstrating absence of bone coverage on sellar floor.

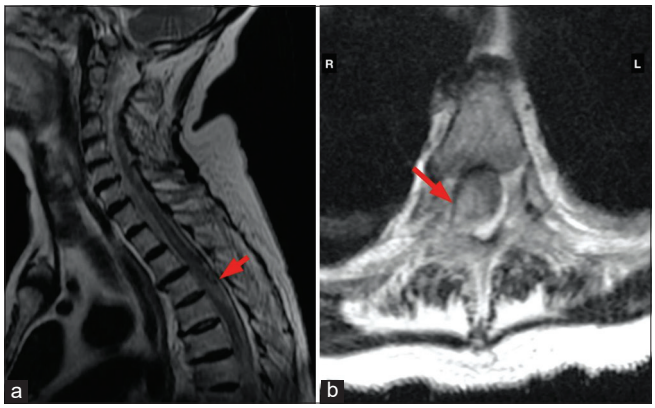

Figure 3: Cervicothoracic MRI. (a) T2-weighted sagittal slice demonstrating hyperintense lesion inside D3-D4 spinal cord. (b) T2-weighted axial slice shows an assymetric hyperintense lesion (right side) pushing spinal contents.

matter hypodensity [Figure 5a-d]. The patient did not have hypertensive episodes and did not use corticosteroid therapy during the postoperative period. Patient and family consent for this manuscript were obtained. This case report was conducted in accordance with ethical standards of the Rio de Janeiro State University and the local ethics committee.

DISCUSSION

RCC

RCC are sellar or suprasellar non-neoplastic cystic lesions arising from remnants of Rathke's pouch.[13,20] The asymptomatic patients can be managed conservatively and observation through serial MRI is appropriate for smaller lesions.[20] When these lesions grow, they can cause mass effect on surrounding structures, such as the pituitary gland, hypothalamus, and optic chiasm and become symptomatic. Surgical intervention is indicated for treatment of headaches, visual disturbances, or endocrinopathies.[12] Reliable surgical approaches have been described for the treatment of RCCs. The current standard is an endonasal, commonly endoscopic, and transsphenoidal approach to access the sellar region.[23] Surgeons commonly utilize this minimally invasive approach to achieve symptom relief, with either GTR of the RCC cyst wall or cyst decompression. Complete cyst excision is less favored than decompression due to increased risk of postoperative complications, particularly endocrinopathies, and intra-operative CSF leak as well as possible injury to the carotid artery or the optic apparatus.[2,5,7,12]

Unfortunately, because decompression alone leaves the cyst wall intact, it has been associated with a higher level of cyst scarring and re-accumulation of the cyst content. Alternative measures have been developed to stabilize the cystic cavity, allowing continual drainage, and avoiding cystic regrowth. One technique that has been popularized is the cyst marsupialization, where the cyst wall is widely opened and the cavity is opened to drain direct into the sphenoid sinus.[12] Marsupialization, also avoids inadvertent injury to surrounding structures when compared to the total resection. By working only on the anterior surface of the cyst, the risk of intraoperative CSF leak is also decreased.[12] In several cases, instead of leaving the marsupialized cyst simply open, a free mucosal graft harvested from the nasal septum, nasal floor, or resected middle turbinate, is placed along the anteroinferior edge of the cyst tract. This provides reinforcement of the draining site and reduces mucosal crusting after surgery. Stenting to open the cavity following marsupialization allows effective treatment, with favorable long-term outcomes.[12] Some external devices such as steroid-eluting stents and silicone stenting, have already been used. However, insertion of a foreign object, may cause trauma to the cyst wall, increase bacterial infections, and foreign body reactions. The use of external devices is a relatively new approach, and further studies are needed to determine long-term outcomes.[12,14] In our particular case, the attempt and pursuit of a GTR favored the occurrence of CSF fistula, which may have contributed to SDAVF rupture.

SDAVF

Dorsal SDAVF (Type I according Di Chiro and Wener[3]) is the most common spinal vascular malformation.[11,16]

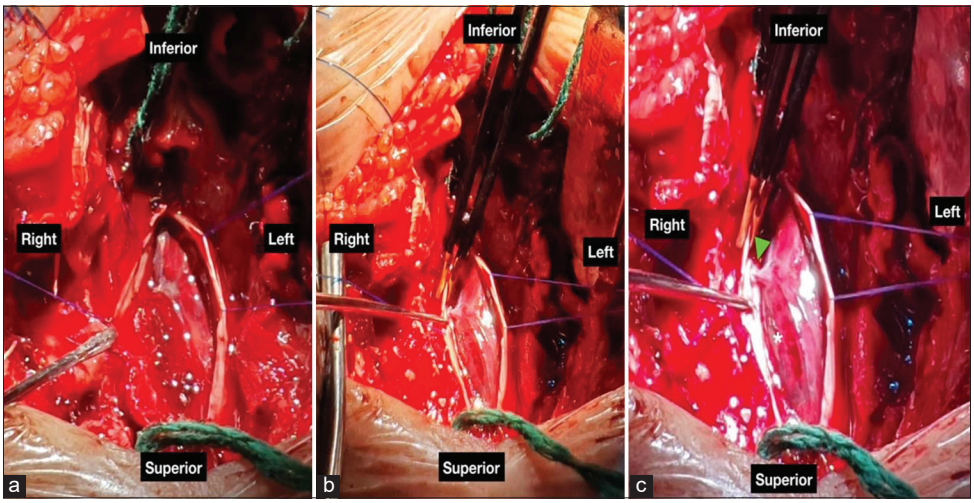

Figure 4: Spinal dural arteriovenous fistula (SDAVF) microsurgery. (a) After dural opening, an important medullary hematoma associated with SDAVF was identified. (b) After blood aspiration, the SDAVF became more evident. (c) The spinal hematoma was aspirated (asterix). After identification of the right T3 artery feeder and venous portion, the shunt was coagulated and cut.

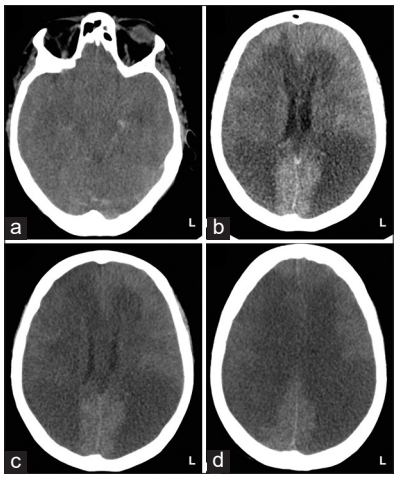

Figure 5: Head CT performed 10 days after SDAVF microsurgery. (a-d) Axial slices demonstrating diffuse white matter hypointensity due to hypoxic encephalopathy.

Patients with SDAVF often present non-specific clinical features that are related to progressive myelopathy. The most common clinical presentations are progressive pain, lower extremity weakness or sensory changes, and sphincter dysfunction.[15] This lesion is composed of a SDAVF in the dura, usually at the dorsal surface of the dural nerve root sleeve in the vertebral foramen, which is fed by a dural artery and drained intrathecally by a medullary vein.[15] This transition is classically located directly underneath the pedicle of the vertebral body, which is supplied by the segmental artery.[11] The intramedullary veins and the radicular vein share a common venous outflow. Then, the increase spinal venous pressure due to arterialization leads to a decreased drainage of normal spinal veins, venous congestion, and intramedullary edema.[11] This congestion, in turn, leads to chronic hypoxia and progressive myelopathy.[11] Direct intraoperative measurement of the vascular pressure of the fistula was found to be as high as 74% of the systemic arterial pressure.[6] This finding may explain why, in some patients, symptoms become worse during physical activity with a concomitant increase in arterial pressure.

Because the lower thoracic region has relatively fewer venous outflow channels compared with the cervical region,[11] the venous congestive edema is likely to be transmitted in a caudocranial direction throughout the spinal cord.[11] This may explain why the first symptoms of myelopathy sometimes reflect dysfunction of the conus medullaris, even though the shunt is located remotely[11] Patients with an SDAVF may show worsening with exercise, upright posture, and Valsalva maneuver. The steroid-induced worsening symptoms are also reported. It has been suggested that steroid-induced worsening may be due to the hypertensive effects of hypercortisolemia.[18]

Diagnosis first relies on MRI that shows central cord hyperintensity on T2-weighted images and increased perimedullary flow voids, most commonly dorsal to the cord.[15] Fast Imaging Employing Steady-state Acquisition sequences have a very high spatial and contrast resolution for intrathecal vessels relative to CSF. Catheter angiography remains the gold standard of diagnosis.[8] Most SDAVFs arise from segmental arteries between T6 and L2.[18] However, our patient had a right T3 fistula with the cord parenchymal abnormalities being most evident from T3 to T4. In that particular situation, as there was an urgency to perform the surgical procedure due to neurological deficit, and angiography would not be available at that time, we opted for emergency spinal decompression.

SDAVFs rarely bleed.[17,18] Subarachnoid hemorrhage and hematomyelia have been reported but are exceedingly rare.[4,10,16] A hemorrhagic presentation is much more common with cord arteriovenous malformations (AVMs).[15] Although the origin of bleeding in SDAVFs is unclear, it is thought that venous hypertension is the cause as it is for progressive myelopathy. Venous hypertension results from arterialized blood entering the medullary vein, thus reaching the valveless coronal venous plexus and radial veins.[11] The formation of a venous varix due to a combination of accelerated venous flow, increased venous pressure, and anatomic changes and fragility of the venous vessel wall can be an important risk factor. These varices may then rupture, causing a hemorrhage. Why these changes occur in some SDAVFs but not in the others remains unknown.[4] The reports[4,11,18] cite the venous hypertension, increased intravascular pressure and venous thrombosis as the main factors to SDAVFs rupture.

In our particular case, these mentioned factors seem to have contributed to SDAVF rupture based on surgical findings (e.g., the single arterialized and thrombosed vein). However, no arterial hypertension episode was noted during the patient's clinical course and corticosteroid therapy was not administrated, which could justify a state of hypercortisolism and SDAVF rupture. In addition, we must consider the fact that there was a sustained CSF fistula with a probable decrease in intracranial and subarachnoid space pressures. The CSF hypotension and subsequent decreased extravascular pressure generating a sudden variation of intra and extravascular pressures (pressure gradient) may also play a key role in the SDAVF bleeding. It is the 1st time that this association is mentioned. Unfortunately, we cannot categorically state whether the SDAVF rupture was due to extravascular factors caused by the CSF fistula or if it happened coincidentally.

Other factors involving SDAVF rupture were also studied by other groups. In a recent review, Yue *et al*.[22] reported three cases of intracranial subarachnoid hemorrhage resulting from non-cervical spinal arteriovenous lesions (i.e.: SDAVF associated aneurysm rupture). Zhu *et al*.[24] demonstrated an increase in annual rate of cerebral AVM hemorrhage during pregnancy and puerperium demonstrating the hormonal influence in arteriovenous lesions rupture. However, this association remains controversial. Vivekanandam *et al*.[21] reported a case series describing an inflammatory CSF examination in patients with progressive myelopathy due to SDAVF. Although there were no cases of SDAVF rupture in this series, the inflammatory process should also be studied and evaluated regarding increased vascular permeability and SDAVF bleeding.

With the evolution of endovascular therapy, catheter-based therapy is often used as the treatment or as a component of treatment of vascular malformations of the spine. Many practitioners advocate endovascular treatment of type I SDAVFs and report good clinical outcomes in some cases.[1] However, certain cases of SDAVFs are not candidates for endovascular treatment: SDAVFs whose arterial feeder originates from the same vessel as a medullary artery, inability to penetrate the SDAVF with glue from the venous side and SDAVF recurrence after embolization. Open surgical treatment of SDAVF has been repeatedly proved safe, effective, durable, and to provide good, reliable clinical outcomes.[15] Regardless of the intervention chosen, whether it is embolization or surgical interruption, the clinical outcome in an SDAVF correlates directly with the patient's preoperative functional status, hence the importance of diagnosis and treatment before symptom progression to a severe stage. The progressive symptoms and occasional rapid deterioration argue against conservative management in all scenarios. The best management of an intradural dorsal SDAVF with hematomyelia and paraplegia seems to be microsurgical resection due to the possibility of decompression and neurological recovery. This includes artery feeding interruption, venous coagulation, cut of the communication and hematoma evacuation.

CONCLUSION

Venous hypertension, venous wall fragility, and venous thrombosis seem to be the main factors involved in SDAVF rupture. In this particular case, reduction of the extravascular pressure and sudden variation in the pressure gradient caused by sustained CSF leak, also appeared to play an important role in SDAVF rupture. It may represent one more complication related to radical resection of RCC.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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