Artículo Original

Hemorragia intraventricular isolada associada à fistula arteriovenosa dural: Relato de caso e revisão de literatura

Isolated intraventricular hemorrhage associated with dural arteriovenous fistula: Case report and review of literature

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ABSTRACT

Dural arteriovenous fistulas are a rare intracranial vascular malformation in the pediatric group. They present as an adult and a pediatric type. A characteristic feature of the pediatric type is the plurality of the fistulous connections and their prevalence in the cavernous sinus. An isolated ventricular hemorrhage has been rarely reported in the literature. Microneurosurgery or endovascular procedures are the treatment options.

We present the case of a child with isolated intraventricular hemorrhage and angiographic study revealing adult pattern of dural arteriovenous fistulae. No treatment was required when subsequent cerebral angiography showed thrombosis of the fistulae. Pertinent literature is reviewed.

KEY WORDS:

Arteriovenous fistulas, dural fistulas, cerebral vascular malformations, intraventricular hemorrhage.

INTRODUCTION

Dural arteriovenous fistulae (DAVF) are uncommon, constituting 10-15% of all cerebrovascular malformations in adults (1,2,3). In the pediatric population these lesions are very rare, representing less than 10% of all intracranial arteriovenous shunts (3,4,5).

From the standpoint of pathology, the feeder arteries are from meninges, the shunt is located in the intracranial duramater and venous drainage is directed to the dural venous sinuses or cortical veins (2).
The DAVF can have different clinical manifestations, including stroke, visual loss, proptosis, transient ischemic attack, seizures and paralysis of cranial nerves (2,4). Although subarachnoid, subdural, parenchymal and intraventricular hemorrhage can occur in different combinations as a complication of a dural fistula, isolated intraventricular hemorrhage was observed in only four cases described in the literature to date.

We report a special case of DAVF with four unusual peculiarities: 1) DAVF in a child, 2) An adult type AVF in the pediatric population, 3) Presentation of an isolated intraventricular hemorrhage, and 4) Spontaneous angiographic thrombosis.

**CASE REPORT**

Ten year old male referred for suspected meningitis, with headache, nuchal rigidity and vomiting for 24 hours. On neurological examination presented with confusion, paralysis of the sixth cranial nerve on the left, meningo-radiccular signs and weak oculo-cephalic reflex. Muscle strength grade IV / V overall, reflexes ++/4 throughout. Coordination and gait were not evaluated due to patient's clinical conditions. Fundoscopic examination showed blurring of the optic disc in the right nasal edge. Cranial CT scan showed isolated bleeding in the fourth ventricle [Fig 1]. Past medical history was pertinent for beta-thalassemia minor since 4 years old.

Cerebral angiography showed a small arteriovenous malformation in the left vermian cerebellar region nourished by branches arising from the posterior inferior cerebellar artery (PICA) draining into the transverse sinus. [Fig 2].

Patient developed supratentorial hydrocephalus by obstruction of the fourth ventricle requiring external ventricular drainage (EVD). After 3 weeks he had clinical improvement with no residual neurological deficit. Control CT scan showed no trace of bleeding or hydrocephalus. He was discharged from the intensive care unit with a plan for definitive endovascular treatment in the immediate future.

Repeat angiography (50 days after the bleeding) showed resolution of the AVF with spontaneous thrombosis of the vascular malformation, eliminating the need for subsequent embolization. On subsequent clinical follow-up three years after discharge the patient remains clinically stable without residual neurological deficits.
DISCUSSION

Vascular lesions composed of abnormal dural arteriovenous connections represent a heterogeneous group of diseases with three distinct pathologic entities: the adult type DAVF; malformations of dural sinus associated with DAVF; multifocal and juvenile DAVF. These last two are unique to pediatric patients (4,6). The DAVF represent approximately 10-15% of all intracranial arteriovenous lesions in adults, the most common locations being the transverse and sigmoid sinuses. In the pediatric population, the descriptions are sporadic, representing less than 10% of all arteriovenous shunts. (7,8,9) The study of Kincaid et al (6) had only two large series published in the literature, containing less than 30 patients. Besides these, only 20 other cases are reported, most demonstrating multifocal DAVF, as is characteristic in the pediatric population. The adult type DAVF can also be found in children, as in the present case. From a clinical perspective, the angioarchitecture and prognosis, DAVF presenting in children are similar to those seen in adults. (5) They generally present with slow flow and are usually located in the cavernous sinus (4). This differs from the present case, in which the DAVF was located in the transverse sinus. The DAVF are acquired lesions that develop after an episode of sino-dural thrombosis and/or venous hypertension, as well as intracranial infections, head trauma, neurosurgery, sino-dural compression by tumors and prothrombotic states. (10) It is presumed that any intracranial pathology leading to sustained venous hypertension can be a predisposing factor for the development of DAVF in an individual with an unfavorable venous anatomy. But the relationship between dural fistulas and venous thrombosis is a two-way street: dural thrombosis can lead to the development of DAVF and vice-versa this can induce sino-dural thrombosis. The clinical manifestations of DAVF (proptosis, conjunctival injection, glaucoma; intracranial: headache, dementia, focal neurological symptoms) are the result of intracranial venous hypertension that develop at a distance from the fistulous point, and do not require the immediate topographical correlation with the DAVF. (2) Intracranial hemorrhage has been reported in up to 35-42% of DAVF, (1,10,11) is believed to be due to disruption of congested cortical veins secondary to retrograde venous drainage (10), and may be intraparenchymal, subdural or subarachnoid. When a patient with a DAVF develops intraventricular hemorrhage it is usually due to the extension of an intraparenchymal hematoma, and was seen in a few isolated cases. (1,10,11) Halbach et al (9) were the first to report DAVF presenting

Figure 1 – CT scan: isolated ventricular hemorrhage in the IV ventricle

Figure 2 - Angiography showing adult type FAVD fed by branches of PICA with drainage to the left transverse sinus
primary intraventricular hemorrhage. Subsequently, only four more cases were reported. (1,8,10,11) The choice of treatment must be based on the natural history of DAVF since the children who survive often have neurological sequela. The treatment should be aggressive in order to cure or at least to exclude cortical venous reflux by either endovascular or microsurgical techniques. Either approach has been reported to be successful, but the choice should be individualized. (6) In the present case spontaneous thrombosis of the vascular malformation waived the original plan. This finding demonstrates the extraordinary dynamic condition of these lesions, in which changes in the pattern of venous drainage can be observed as a result of thrombosis of the vein or dural sinus that drain the fistula. The DAVF can thus progress or undergo spontaneous occlusion of the draining veins or by dural sinus thrombosis. Progression in grade occurs in less than 5% of the total DAVF, and total spontaneous thrombosis is rare. (2) The disappearance of an intracranial arteriovenous malformation has been hypothetically attributed to thrombosis following an episode of hemorrhage. (7) The remission of the vascular anomaly in this patient was observed after almost two months of initial angiography, occurring earlier than previously described in the literature. Clinical follow-up with periodic examinations is justified, because angiographic non visualization of the DAVF, does not mean it has completely disappeared. There are several reports of angiographically occult AVF, including cases where there was disappearance and reappearance in sequential angiography. Thus, the mere absence of an AVF in an angiogram does not exclude the presence of a potentially dangerous AVF, requiring long-term monitoring. Although rare, DAVF are cerebrovascular malformations that should enter the differential diagnosis of children with ischemic or hemorrhagic stroke, seizures or focal neurological deficits. The typical pattern of pediatric DAVF is multifocality, but can be found also as adult type DAVF in children. The underlying pathophysiologic mechanism is related to any condition that promotes prolonged venous hypertension. Clinical manifestations are due to intracranial hypertension resulting from venous stasis and/or hemorrhage secondary to the venous engorgement. Treatment should aim to cure the DAVF, and can be either microsurgical or endovascular. Angiographic follow-up should be maintained even after an angiography demonstrated disappearance of DAVF, either after treatment or spontaneous thrombosis as there are many cases reported of recurrence.

REFERENCES


