Intracranial dural arteriovenous fistula. A case report

Alexis Román MatusM.D.
Jesus Manuel Higuera Cardenas M.D.
Juan Daniel Naranjo Hernandez M.D.
Carlos Ivan Amezcua Equihua M.D.
Marianna Carlos Felix M.D.

Tijuana, Mexico

Case Report
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Background: Dural arteriovenous fistulas (DAVF) are abnormal connections between arteries and dural veins, or venous sinuses originated from dural folds. Most common etiology is idiopathic. They are one of the most common non-traumatic causes of intracerebral hemorrhage in patients of 45 years old and younger. The gold standard for diagnosis is angiography by intraarterial catheter. The actual surgical management involves the disconnection of venous drainage from the fistula.
Clinical case: A 7-year-old girl comes to emergency service presenting holocraneal headache of sudden onset, 10/10 intensity, projectile vomiting and loss of consciousness with tonic-clonic seizures and sphincter relaxation. Computed angiotomography with 3D reconstruction showed presence of right frontal parenchymatous bleeding with intraventricular invasion, secondary to arteriovenous malformation. Surgical procedure was performed with disconnection of venous drainage. Reevaluation after thirty days of hospital discharge showed remarkable clinical improvement.
Conclusions: Disconnection of fistula is enough as treatment for most cases of DAVF.

Key words: Vascular fistula, arteriovenous fistula, venous sinus

Introduction

Dural arteriovenous fistulas (DAVF) are anomaly connections between dural arteries and veins, or venous sinus originating in the dural folds. The most common etiology it's the idiopathic one. The DAVF are the most common causes of non-traumatic brain hemorrhage in age equal or less to 45 years.[1] With frequency they are located proximus or related to the dural venous sinus wall that is occluded or stenosed. Imaging studies such as the magnetic resonance image (MRI) and computed tomography (CT) are usually normal in DAVF. The intra-arterial catheter angiography continues to be the gold standard for imaging study of DAVF.[2] Between the management options trans-arterial and trans-venous embolization, stereotactic radiosurgery, as well as open surgery are included in the management options. We report a new case and compare it with what was previously reported.[3]

Case report

A 7 year old female was referred to the neurosurgery service from the health center as she presented sudden alert status deterioration, she had no significant medical history for the described condition. She starts on august 29th 2018 as she presents sudden holocranial headache, 10/10 intensity, followed by large projectile vomiting and loss of alert status united to tonic- clonic seizures with sphincter relaxation for approximately 20 minutes. As she arrives to the hospital, a simple cranial tomography was performed (figure 1) where a hyperdense collection towards the right frontal region was found (.3 cm from the internal table) with an approximate 31 cc volume, furrows of both hemispheres, conditioning a mass effect and deviating from midline .7 cm to the left. It was also observed the presence of a hyperdense material (72UH) at an supra and infratentorial level of the ventricular system. She was admitted to the pediatric intensive care unit (PICU) where she was kept under sedation and anti-edema measures. A cranial computed angiotomography (figure 2) and 3D reconstruction was performed where a parenchymatous bleeding was found of right frontal localization which measured 4.1 cm sagittal x 3 cm transversal x 4.7 cm anteroposterior. Said secondary hematoma to an arteriovenous malformation, which measured by tomography less than 3 cm (1 cm x 1.2 cm), an eloquent area was found and without drainage to the deep venous system, so it is cataloged with a Spetzler score 1. The presence of an anomalous vessel was evident, ecstatic and dolic, giving the appearance of being originated from the dura.

A Falconer type incision was performed, mucocutaneous dieresis with a pericranial flap. A frontotemporal craniotomy was performed with a

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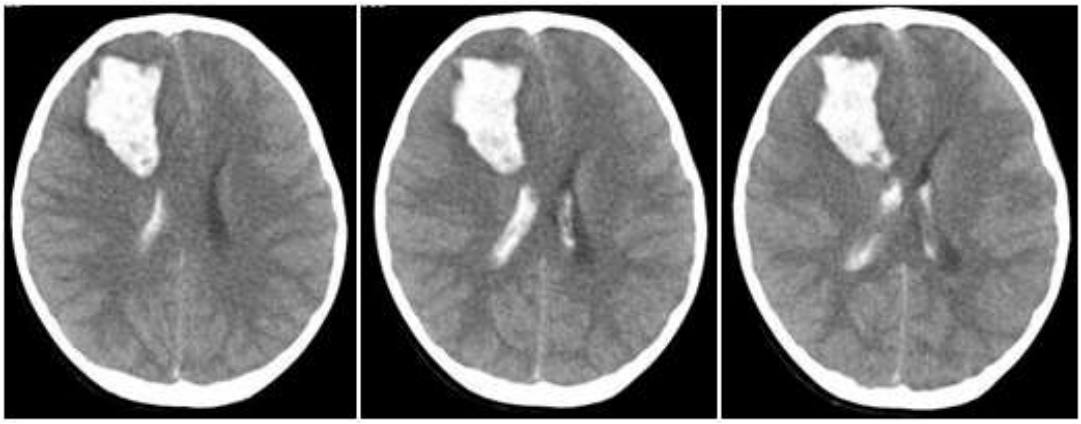

Figure 1. Cranial CT scan. Axial.

pneumatic craniotome, C durotomy, and a severe cerebral edema with a transcalvarial herniation was found. When the durotomy was made a dural drainage vein was found to be communicated with the leptomeningeal veins. The venular bed is located and surrounded (Figure 4), the principal drainage vein is coagulated and the intraparenchymal hematoma is subsequently drained. Duralplasty was performed and plane closure.

Findings: Dural arteriovenous fistula with a small caliber principal drainage vein, as well as bed of arterialized veins, with a parenchymal hematoma of 30 cc, bleeding of 100 cc, no accidents or incidents, right pupil of 4 mm, left of 2 mm.

After 2 weeks of postoperative surveillance in PICU, she evolves favorably and her discharge is decided, highlighting function in cognitive functions and weakness in four extremities, upper limbs right and left with muscular strength ⅗ and ⅖ in Daniel's scale, respectively. Lower limbs right and left with muscular strength ⅗ and ⅕ in Daniel's scale, respectively.

She was evaluated three months later by the neurosurgery service with a simple cranial tomography (Figure 5) and a control cranial angiotomography (Figure 6), finding no evidence of bleeding. The patient was found to have clinical improvement, language improvement, mobilization of the four extremities, right thoracic limb 5/5 , left thoracic limb ⅘, right lower limb ⅘, left lower limb ⅗ in Daniel/s scale, surgical wounds without complication signs.

Discussion

The DAVF are defined as aberrant connections between dural artery and dural venous sinus or a cortical vein. The DAVF are presented with more frequency in patients between the fifth and sixth decade of life, representing about 10-15% of the intracranial arteriovenous malformations in adults, being its percentage a little higher in posterior fossa (35%).[4] In the case of the pediatric population, the cerebral arteriovenous malformations are the most

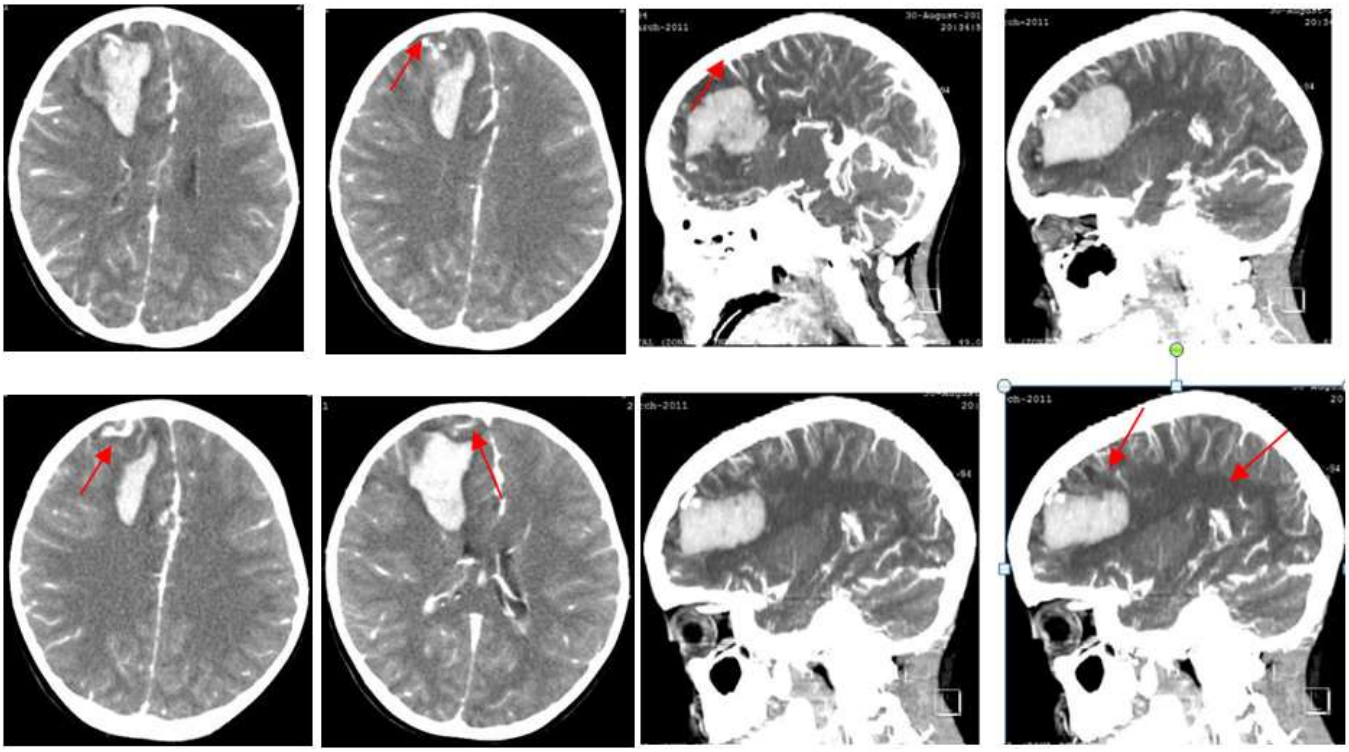

Figure 2. A. Cranial angio-CT scan (Axial). B.Cranial angio-CT scan (Sagittal).

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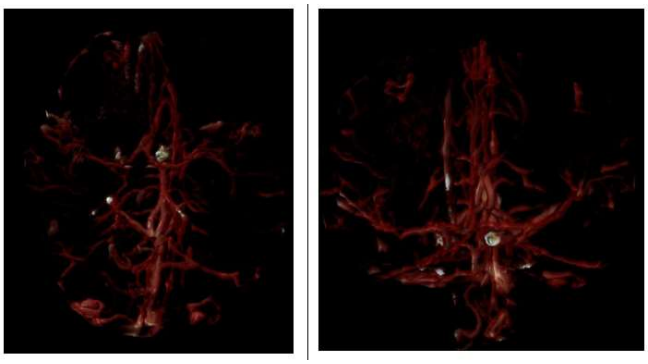

Figure 3. Cranial angiotomography. 3D Rconstruction.

common cause of intracranial hemorrhage, showing up 50% of the time, nevertheless, DAVF are only the 10% of the mentioned malformations. [5]

As a risk factor for their formation, we find chromosomal anomalies like hereditary hemorrhagic telangiectasia syndrome and type 1 neurofibromatosis. Other risk factors are hypercoagulability (being risk factor sinus thrombosis and subsequently DAVF formation) such as leukemia and smoking, and head trauma.[5,6]

The symptoms vary depending the location of the injury (being most frequent the cavernous sinus, cribriform plate, transverse sinus, sigmoid and tentorium in adults; in the pediatric population, is the torcula, superior sagittal sinus, transverse sinus and cavernous sinus) and its corresponding venous drainage, in this case, DAVF was found in the frontal region and the nutrient artery was the anterior ethmoidal; the symptomatic vary between asymptomatic, cognitive impairment, intracranial hemorrhage and sudden death.[7] It has been reported that being female is an independent risk factor as a predictor of hemorrhage, which is consistent with the sex of this case. It has been seen that the DAVF that are farthest from the torcula, theoretically should have more favorable results, as there is at least one normal cerebral venous sinus for drainage.[8]

In the pediatric population, the presentation tends to vary with the presentation age, being the most frequent, heart failure, respiratory distress, hidrocephalus in patients less than 1 year old; while that in the older than 1 year is more common the presence of focal neurologic deficits.[9] In his long-term experience in pediatric DAVF, Lasjaunias shows that this can be lethal up to 10 years after presentation. [10]
It is important to highlight that patients with DAVF presenting in childhood are more prone to have novo formations of DAVF lately in their life.[5,11]

The angiographic features suggestive of increased bleeding risk include the localization in the petrosa or the straight sinus, the presence of leptomeningeal venous drainage and the presence of varicose veins in the drainage vein. [12] It was decided to perform the surgery when having the computed angiotomography with 3D reconstruction, because it shows a sensibility and specificity of 94% and 93% respectively for the detection of these lesions.[13]
The DAVF with cortical venous drainage (Borden type II-III) usually have a more aggressive clinical course. In a series of 20 patients with DAVF with initial presentation of bleeding, a rebleeding rate of 35% was found between the first two weeks.[14]

The intracranial hemorrhage, the focal neurologic deficit or death, were found in 2% of the DAVF Borden type 1, 39% in type II and 79% in type III, being the last one which had the patient. [15] It's important to point out that in this case the fistula was found in the anterior part of the sagittal sinus, compared with other cases where the fistula is found in the posterior part of the sagittal sinus.[16]

According to the evidence reported until now, it has been proved that opting for less invasive procedures (closure of nutritional vessel of the arteriovenous fistula vs fistulotomy) results in a lower risk of intracerebral hemorrhage, neurologic deficit and morbimortality in general.[17]

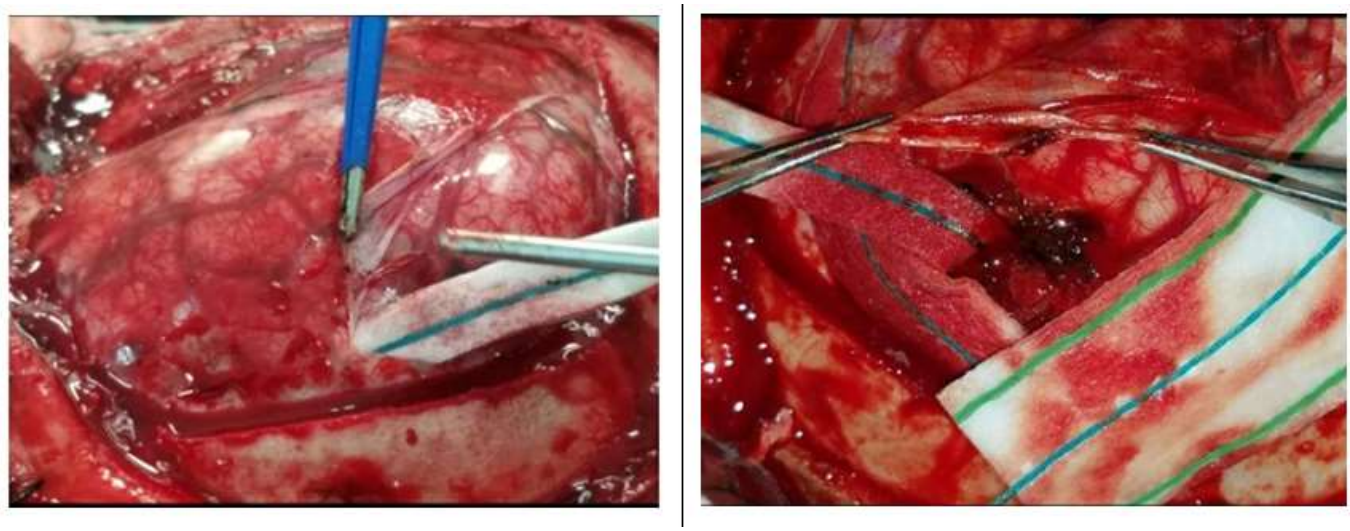

Figure 4. Transoperatory findings.

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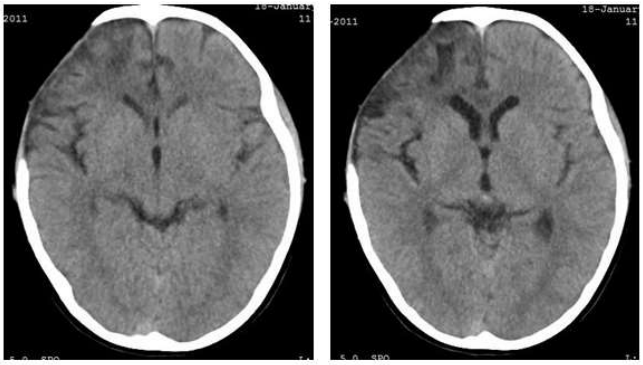

Figure 5. Cranial CT scan. Control

Although previous surgical treatments implied complete surgical resection of the fistula, this technique has been abandoned in the last years due to recognition that the sole disconnection of the venous drainages of the fistula is curative, being this last what was done in this case.[18] This strategy helps to reduce the risk of the fistula, including intracerebral bleeding and neurologic deficit.[19] The cortical venous drainage disconnection is a more simple procedure than the complete cleavage of the fistula, and takes a lot less risk of morbidity. [19,20] The procedure involves the exposition of the fistula and coagulation of any found nutrient artery, the arterialized veins are exposed and coagulated as close to the fistula as possible, the normal veins should be preserved, previous steps performed in this case.[20] The surgical treatment for the DAVF with venous reflux has been associated with an acceptable angiographic cure rate.[21] Previous approaches were quite extensive and require the coagulation of nutrient arteries, cleavage of the dural sheet, arterialized leptomeningeal veins and the possible sacrifice of the dural venous sinus, associated with a high rate of mortality and morbidity, being the principal complication bleeding at any time in the surgery.[22]

The intracerebral Hemorrhage score to assess mortality at 30 days in intracerebral hemorrhage shows a score 4 carries a 30-day mortality of 97% with characteristics similar of those of our patient, nevertheless, in this case it has been demonstrated an adequate clinical evolution at 30 postoperative days.[23] Said scale decided not to be used for making therapeutic decisions due to population size used for the making of the score, lack of stratification of patients according to their age and morbidities.

There is currently no prognostic assessment tool for intracerebral bleeding in the pediatric population.

The classic methods for treatment are being rethought for the treatment of cerebrovascular lesions.

Conclusions

Currently it is sufficient to disconnect the venous drainage in the DAVF for the treatment of this pathology, which can be made in second level hospitals as long as you count with the medical equipment and technical neurosurgical team available.

Conflicts of interests

The authors declare that there are no conflicts of interest

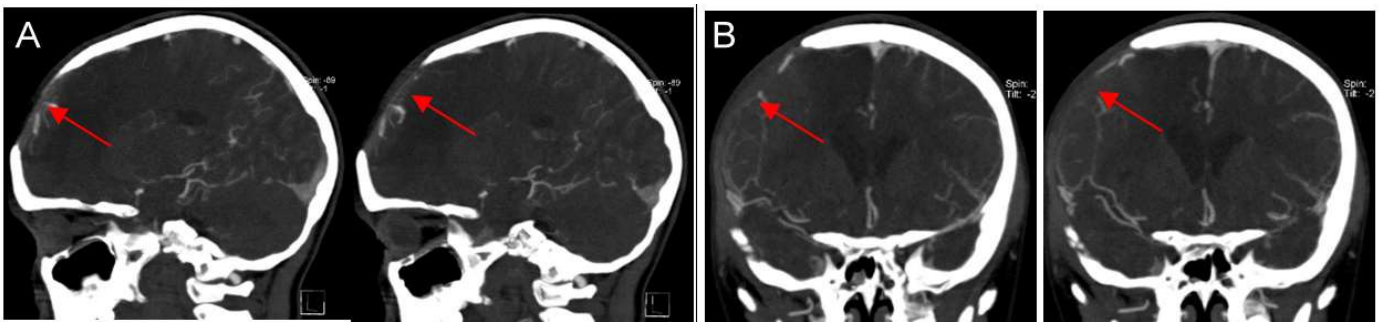

Figure 6. Cranial angiotomography control. A. Sagittal. B. Coronal. Red arrows show absence of vein drainage.

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Alexis Román Matus
Department of General Surgery
Instituto Mexicano del Seguro Social
Hospital General Regonal No. 1
Tijuana, Mexico
alexisrm188@gmail.com

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