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CASE REPORT

Giant dural arteriovenous fistula in a pediatric patient: positive outcome following surgical treatment

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Abstract

Purpose Our purpose is to present an atypical case of a 4-month-old patient with a giant dural arteriovenous fistula (DAVF).
Methods Presentation of a case report and review of the literature.
Results The DAVF arterial supply was through the middle meningeal artery bilaterally and the anterior and middle cerebral arteries on the right hemisphere. The venous drainage was through the posterior two-thirds of the superior sagittal sinus. The endovascular team performed an embolization to reduce the flow of the lesion, and finally, the surgical team completed the excision of the residual venous sac, without causing any significant neurological deficit. We used a double surgical approach done with two surgical teams in order to optimize the hemostasis control and reduce morbidity and mortality.
Conclusion Midline DAVF usually has devastating consequences in children. Endovascular treatment is the first choice since it has lower mortality. Nevertheless, it requires multiple interventions, and the cure of the disease may not be achieved. We believe that joint endovascular and surgical treatment, supported by a reliable multidisciplinary medical team, is a good option for this type of lesions.

Keywords Dural arteriovenous fistula · Vascular malformation · Surgery · Endovascular · Pediatrics

Introduction

Dural arteriovenous fistulas (DAVF) are abnormal communications between dural arteries and veins. The venous drainage could be through a cortical vein or it could go directly into the venous sinuses, causing dilation and thrombosis [1]. The exact prevalence of these lesions is unknown; Garcia-Monaco [2] had reported that DAVF account for 10% of all intracranial arteriovenous shunts in children.

Lasjaunias [3] had described two types of DAVF, those who drain into the lateral sinus, with better prognosis, and those who drain into the superior sagittal sinus or to the torcula, which usually evolve poorly.

The preferred treatment is embolization due to its lower mortality; however, it requires multiple interventions and in most cases the lesion relapses. On the other hand, surgical treatment is generally definitive, but it carries higher risks in non-experienced hands. Mortality for this type of vascular lesions was described as 38% in children and 67% in neonates [4]. If the torcula had been involved, the result was unfavorable in 71.4% of cases [1].

We presented a 4-month-old patient with a giant DAVF with superior sagittal sinus drainage. The lesion had been embolized and then resected with good postoperative results and minimal morbidity. To our knowledge, this is one of the few cases of a giant DAVF with superior sagittal sinus drainage and successful treatment by a surgical approach.

Case report

Background

A 4-month-old female patient was referred to our outpatient clinic by her mother because she had had, since birth, a bulging anterior fontanelle and macrocephaly. The physical exam had shown normal neurological development, but a bulging tumor was palpable in the anterior fontanelle. She had a cephalic perimeter of 42 cm which was more than two standard

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deviations above average. An echocardiogram had shown early signs of high-output cardiac failure and cardiomegaly. The ophthalmologic exam was normal, and the lab results had shown anemia and coagulation disorders. After the exams were finished, the patient was medicated with furosemide and phenobarbital.

Preoperative images and vascular intervention

We performed a computer tomographic angiography (CTA) which revealed a giant DAVF occupying approximately the anterior two-thirds of the cranial cavity. The lesion had spontaneous hyperdense areas and heterogeneous enhancement after contrast. Some of the hyperdense areas could be interpreted as thrombosis or hemorrhage. On the CTA reconstruction, we can appreciate multiple venous aneurysms, with drainage to the superior sagittal sinus. The lesion had caused an important mass effect in the surrounding structures. The superior sagittal sinus had marked dilation of the posterior two-thirds of the superior sagittal sinus (SSS), torcula, straight sinus, and the vein of Galen (Fig. 1).

Digital subtraction angiography (DSA) was performed showing arteriovenous (AV) shunts in the wall of the dural sinus malformation (DSM) fed by the middle meningeal artery and the superior temporal artery, branches of the external carotid artery (Fig. 2a), and also by the right middle cerebral artery, branch of the internal carotid artery. Both middle meningeal arteries were embolized at the fistulous sites using glue. Control DSA after embolization compartments of the DSM are clearly depicted (Fig. 2b–c)

The child had returned 1 month after the procedure with a bulging fontanel and vomits. A new magnetic resonance (MR) had shown that the DAVF was partially thrombosed with even more edema, midline shift, and compression of the surrounding structures. The image had also revealed a subacute intraparenchymal hemorrhage in the insular cortex. The venous phase had shown that the two-thirds of the sagittal sinus were patent (Fig. 3a–c)

Surgery

The surgery had been planned 2 days after the last endovascular treatment. The patient was 5 months at the time of the surgery. A team was designed especially for this surgery formed by neurosurgeons, neuroradiologist, pediatrician, and anesthesiologist. We had performed a simulated surgery

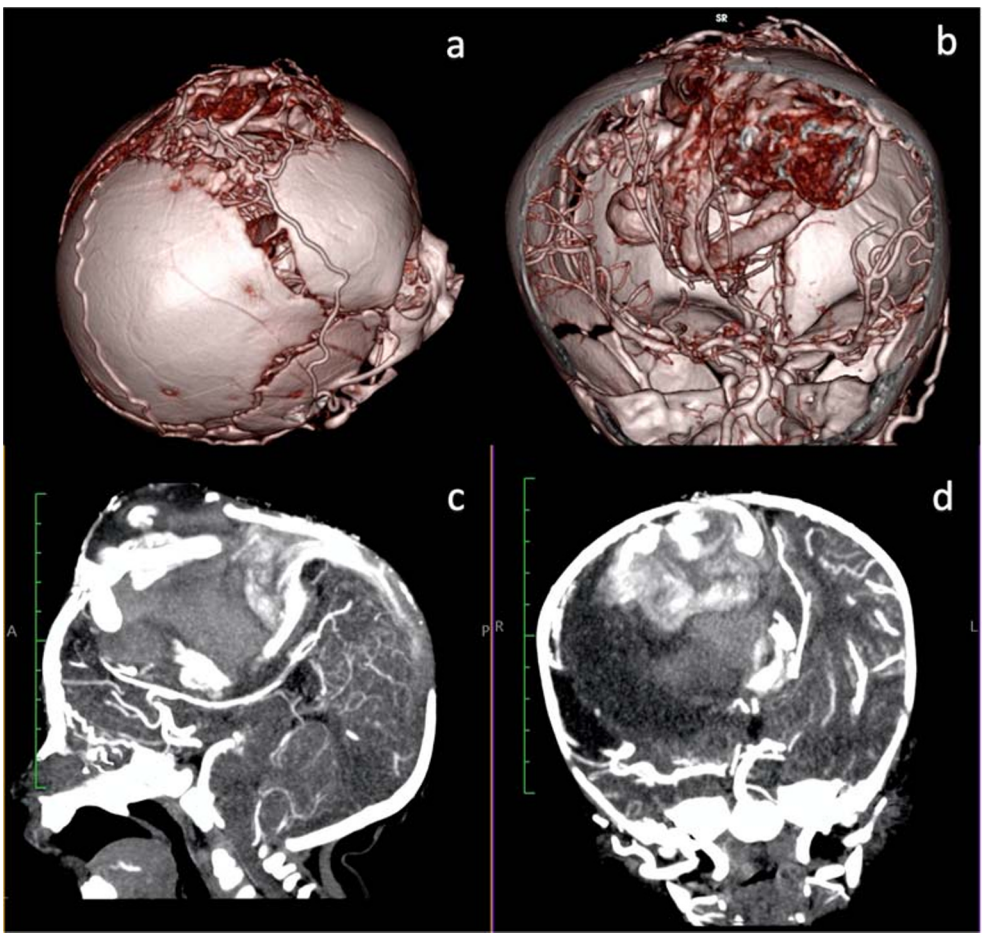

Fig. 1 Computed tomography angiography (CTA) with 3D reconstruction. **a** 3D reconstruction of the brain CTA showing the dilated middle meningeal artery and the occipital artery nourishing the dural arteriovenous fistula (DAVF) through an enlarged anterior fontanelle. **b** Posterior view of the CTA 3D reconstruction where we can see the branches of the middle and anterior cerebral artery feeding the DAVF and dilatation of the superior sagittal sinus. **c** Sagittal view of the CTA showing a large extra-axial lesion over the frontal convexity with heterogeneous attenuation probable due to thrombosis and hemorrhage. The remaining superior sagittal sinus is also dilated. **d** Coronal view of the CTA showing the right-sided lateralized mass with midline shift and the mass effect on the surrounding structures

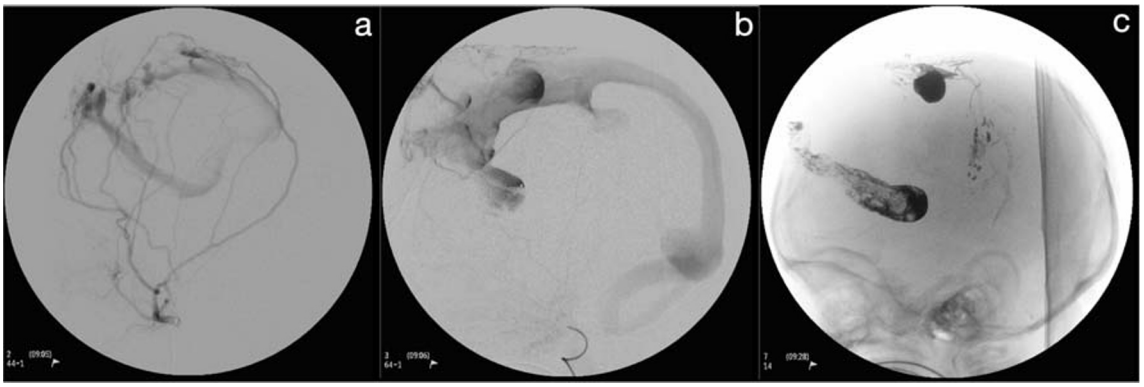

Fig. 2 Digital subtraction angiography (DA). **a** DSA showing arteriovenous (AV) shunts in the wall of the dural sinus malformation (DSM) fed by middle meningeal artery (MMA) and superior temporal artery branches. **b** Late phase of DSA. The compartments of the DSM involving the anterior part of the superior sagittal sinus are evident. **c** Partial embolization with glue of the AV shunts in the wall of the DSM filling its compartments

reviewing each step carefully, we studied the arterial afferences and the venous outflow that we were going to encounter during surgery, and we planned ahead how to manage them. We had also discussed the dural and calvarial reconstruction after the DAVF was removed. We decided to make two teams at each side of the patient. Each team was composed by two neurosurgeons, two scrub nurses and one anesthesiologist.

The surgery began with the delimitation of the superficial temporal arteries with ultrasound (Fig. 3d). A biauricular incision and a subgaleal dissection were performed to expose the afferences from the superficial temporal arteries. Once the arteries had been identified, they were properly managed with the bipolar coagulator. After we had gotten control of the first layer of arterial afferences, we started the bifrontal-parietal craniotomy. Once we had removed the bone, the bilateral middle meningeal arteries supply was revealed. The arteries were also coagulated in the surface of the dura mater with bipolar. We opened the dura with ultrasound assistance to avoid damage to any vascular structure. Once we had exposed the lesion, we dissected the normal parenchyma from the lesion, and we ligated and cut the anterior insertion of the superior sagittal sinus to allow free movement of the sac. We identified the anterior and middle cerebral arteries afferences coming from beneath the

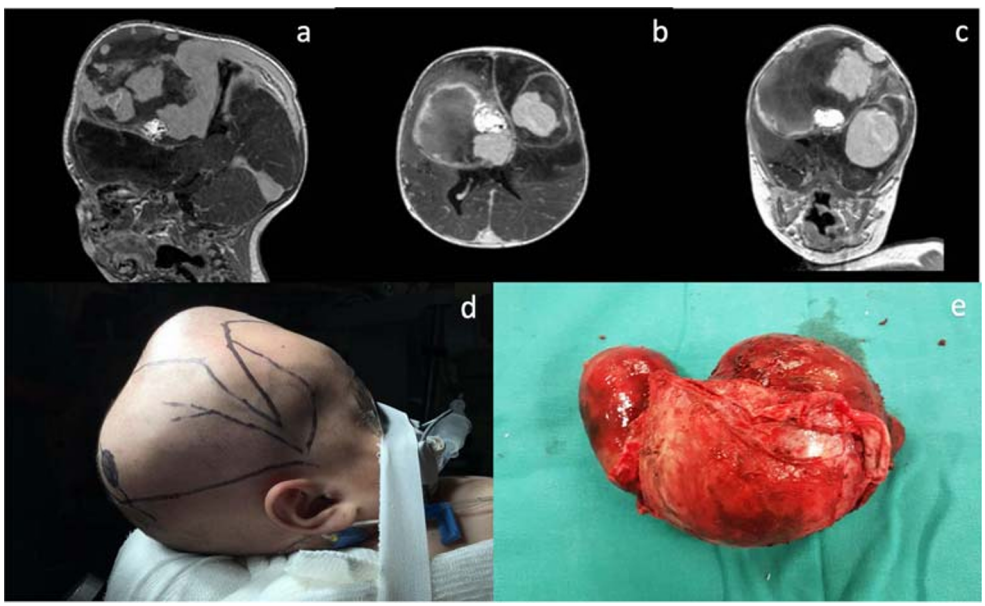

Fig. 3 Magnetic resonance imaging (MRI) of the brain. **a** Sagittal view of the brain MRI after the first embolization, partially thrombosed venous sacs and hemorrhage can be seen at different stages. Flow is observed in the posterior two-thirds of the superior sagittal sinus. **b** Axial view of the brain MRI showing the lesion with different heterogeneous areas of thrombosis and hemorrhage. Also, we can see the embolization material. **c** Coronal view of the brain MRI showing a hyperintense area corresponding to subacute bleeding in the left insula. Intraoperative images. **d** Intraoperative image of the patient in position for surgery with the temporal superficial artery marked with ultrasound. **e** Final image of the dural arteriovenous fistula after resection

lesion; they had been partially embolized, so we put a vascular clip and coagulated the entry point.

After the sac had been completely dissected, and all the arterial afferences had been controlled, we put a vascular clamp in the middle of the superior sagittal sinus, right after where the sac ended. Once the sagittal sinus had been ligated and cut, we removed the lesion from the skull without any complications (Fig. 3e).

To get a watertight dural closure, we used pericardium. For calvaria reconstruction, we used the patient's bone and we fix them with microplates and screws.

Postoperative period

The patient remained in the intensive care unit, requiring ventilatory mechanical assistance for 48 h, after that she was extubated without difficulty. Despite the extensive resection of the lesion, she had no neurological deficit nor developed hydrocephalus.

A control CT scan was performed that showed complete excision of DAF and the anterior third of the superior sagittal sinus. In addition, the CT had shown the remains of the embolization material in the afferent arteries and leukomalacia in the frontal lobes, which was already observed in the previous images (Fig. 4).

Currently the patient is 2 years old, the physical examination shows developmental milestones according to age, and the physical examination reveals a left upper limb paresis with spasticity predominantly in the hand and the rest of the extremities have preserved motor strength. The ophthalmological examination continues to be normal. She does not take any medication.

Discussion

The exact mechanism resulting in the formation of dural sinus malformations remains unclear. Okudera et al. [5] have suggested that it is the result of the ballooning (or enlargement) of the transverse sinus at 4–7 months of intrauterine life due to the lack of formation of the jugular sinus. This physiological mechanism usually reverses after 2 years once the jugular sinus is channeled. The authors suggest that in these patients, by some unknown mechanism, the transverse sinus is not remodeled and produces a retrograde dilation of the venous drainage. This theory could explain the dural sinus

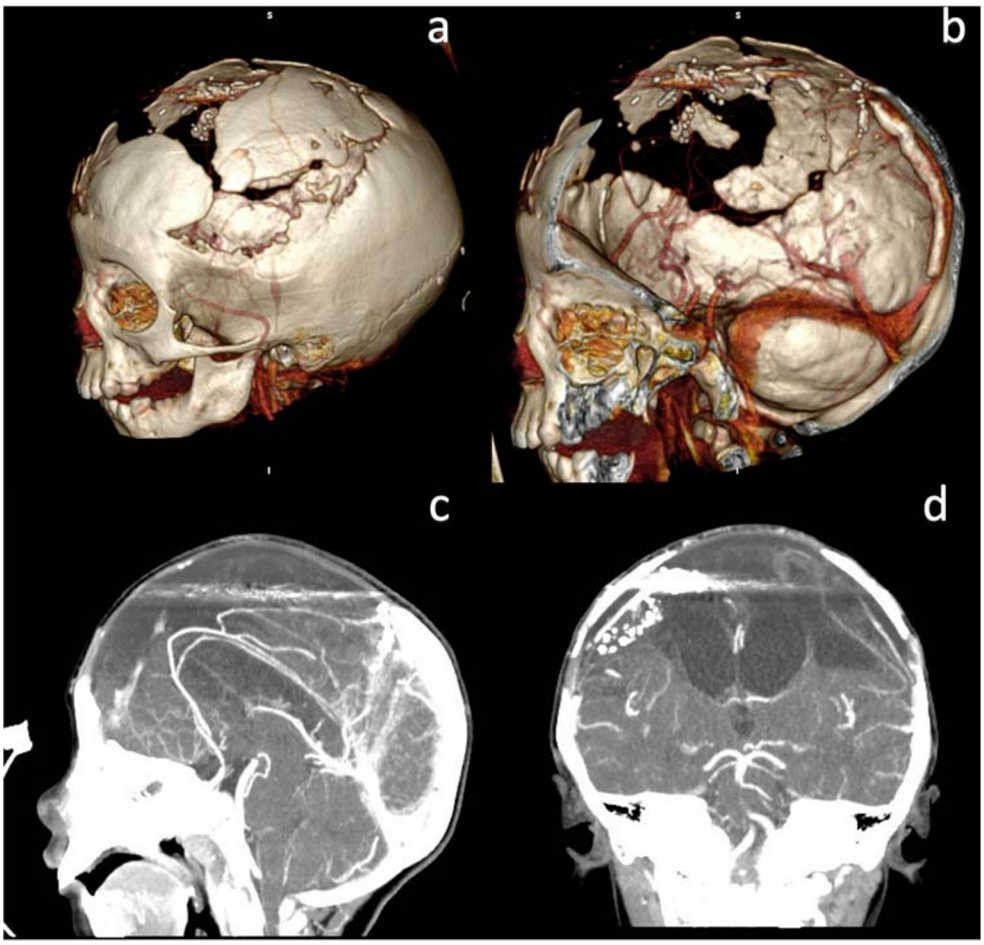

Fig. 4 Postoperative brain computed tomography angiography (CTA). **a** External 3D reconstruction of the CTA showing the resolution of the afferent branches of the external carotid artery. **b** Internal 3D reconstruction of the CTA showing the reduction of the arterial afferent branches of the internal carotid artery and the resection of the two anterior thirds of the superior sagittal sinus. **c** Sagittal view of the CTA showing complete excision of the lesion together with the anterior two thirds of the superior sagittal sinus. In addition, we can see that the anterior cerebral artery recovered its usual position, as well as deep venous drainage. **d** Coronal view of the CTA showing a slight ventricular dilatation with leukomalacia of the cerebral parenchyma

malformation, but this mechanism could not explain the appearance of the arteriovenous fistula in the sinus wall.

Two hypotheses address the pathogenesis of the DAVF. One is that the fistula arises from "latent" channels between the external carotid artery and the venous channels within the dura, opening in response to venous hypertension. The other suggests that the new vascular channels are stimulated by angiogenic factors arising directly from sinus thrombosis or as a result of tissue hypoxia.

Lasjaunias et al. [3] were the first group to report a series of cases of dural venous malformations in children. They classified the malformations into three types: dural sinus malformations (DSM), dural arteriovenous shunts of the infantile type, and dural arteriovenous shunt of the adult type. The DSM are subclassified into two types according to the location of the fistula: jugular bulb with high-flow fistula (usually with a good prognosis as they maintain the contralateral venous drainage) and posterior sinuses (which may or may not involve the torcula) with slow-flow fistula (with poor prognosis due to the presence of great venous dilation and spontaneous thrombosis that generates venous infarcts and hemorrhage).

The symptoms will depend on the size of the lesion and its location. Larger lesions involving the midline sinuses usually generate symptoms before the year of age, such as cardiac overload, respiratory distress, and hydrocephalus. Smaller or lateral lesions usually produce symptoms after 1 year of life, such as focal deficits, macrocephaly, headache or cognitive deficits.

Mortality for this type of injury is 38% in children and 67% in neonates [4]. If the torcula is involved, the result is unfavorable in 71.4% of cases. The management of DAVF with sinus thrombosis is poorly documented due to the rarity of these lesions [6].

Because of its lower mortality, the endovascular approach is the treatment of choice [7]. However, one should consider that in most cases multiple interventions are required to achieve total occlusion of the fistula and the disease-free survival to be as low as 30% [4, 8–10]. In addition, incomplete embolization can lead to other complications such as the eventual development of cavernous malformations secondary to venous congestion due to the dural arteriovenous fistula [10–13].

Surgery is a valid option; however, it is not the treatment of choice due to its high morbidity and mortality. Our team has experience in the surgical treatment of these lesions; however, we had not yet found a midline DAF comparable with the one described in this case [14, 15]. The benefit of the surgery is that, if the lesion is completely resected, the patient is cured. But, it could lead to multiple complications in unexperienced hands [16].

The case presented by the authors has the peculiarity of having several adverse prognostic factors like midline location of the fistula with thrombosis, infarction, and hemorrhage, in a patient younger than 6 months. We decided to take a surgical approach because the previous embolizations did not prove to be effective in treating the lesion. Also, we had a multidisciplinary team and experienced neurosurgeons to try a surgical approach. The radiologic department performed the endovascular pre-surgery embolization to reduce the high flow of the fistula, and the neurosurgery department performed the surgical resection with two groups of neurosurgeons and anesthetists who were working together to achieve the excision without significant complications.

We believe that in experienced hands and with the help of a dedicated team the surgical excision of these lesions could be performed safely.

Conclusion

Midline DAVF usually has devastating consequences in children.

Endovascular treatment is the first choice since it has lower mortality. Nevertheless, it requires multiple interventions, and the cure of the disease may not be achieved.

We believe that joint endovascular and surgical treatment, supported by a reliable multidisciplinary medical team, is a good option for this type of injury.

Code availability Not applicable

Authors' contribution All authors contributed to the study conception and design. The first draft of the manuscript was written by Amparo Saenz and Beatriz Mantese, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript

Availability of data and material Not applicable.

Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflict of interest.

Ethical approval This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the Ethics Committee of Garrahan's Pediatric Hospital.

Consent to participate Informed consent was obtained from the parents of the participant included in the study

Consent to publish The parents of the patient signed an informed consent regarding publishing their data and photographs.

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