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CASE REPORT

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Endovascular treatment of a cerebral single-hole arteriovenous fistula associated with a giant aneurysmal sac in a pediatric patient

Abstract: Cerebral arteriovenous fistulas (AVFs) are rare in childhood and are generally associated with arteriovenous malformations (AVMs). They may lead to an increase in intracerebral pressure, seizure, hemorrhage, or cardiac decompression. They may be treated with endovascular embolization. Herein we present the radiological findings, before and after endovascular treatment, of a pediatric patient with a cerebral AVF associated with a giant aneurysmal sac.

Key Words: Arteriovenous fistula, cerebral arteriovenous malformation, cerebral MRI, DSA, endovascular treatment, venous aneurysm

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Dev anevrizmatik kesenin eşlik ettiği serebral arteriovenöz fistülü olan pediatrik olgunun endovasküler tedavisi

Özet: Serebral AVF'ler çocuklarda nadir görülen, genellikle AVM'lar ile birlikte olan hastalıklardır. İntrakraniyal basınç artışı, nöbet, hemoraji, kardiyak dekompresyona neden olabilirler. Endovasküler embolizasyon ile tedavi edilebilirler. Bu yazıda, dev anevrizmatik kesenin eşlik ettiği serebral AVF' ü olan pediatrik bir hastanın, tedavi öncesi ve endovasküler tedavi sonrası radyolojik bulguları sunulmaktadır.

Anahtar Sözcükler: Serebral arteriovenöz fistül, serebral arteriovenöz malformasyon, MRG, DSA, endovasküler tedavi, venöz anevrizma

Introduction

Arteriovenous malformations (AVMs) are the most common lesions of the cerebrovascular system in children (1,2). In contrast to adult cases, in children they are 10 times more frequent than cerebral aneurysms (1). AVMs are congenital lesions located between arterial and venous circulation that arise due to developmental anomalies of the arteriolar-capillary network. On the other hand, arteriovenous fistulas (AVFs) are acquired or congenital lesions characterized by abnormal connection between the arterial and venous systems, but without the capillary bed or nidus observed in AVMs (3,4). AVFs are commonly associated with giant venous sacs (1).

AVFs rarely are asymptomatic. They commonly present with an increase in intracranial pressure, seizure, cerebral hemorrhage, and cardiac decompression findings, which necessitate treatment (1,3). They may be treated surgically or endovascularly (3).

Herein various radiological findings of a pediatric patient with a cerebral AVF associated with a giant aneurysmal sac, before and after endovascular treatment, are presented.

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Case Report

A 10-year-old female patient presented with complaints of headache, behavioral changes, and urinary incontinence that began approximately 2 years earlier, perioral seizures that began 2 months earlier, and an inability to hold her breath during the previous 3 weeks. The patient was referred to our hospital following a cranial MRI evaluation by a pediatrician at another institution that showed an aneurysm.

Her history revealed that she had her first seizure when she was 7 years old, at which time she experienced trembling, with eyes focused on a particular point. She had a fever (38 °C) at the time of the seizure. The duration of the seizure was unknown. She was diagnosed as having had a convulsion due to high fever and was treated symptomatically, without medication.

When she was admitted to our hospital her physical findings were normal. There was indistinct left central facial paralysis in her neurological examination. Her body temperature, blood pressure, pulse, and respiratory rate were within normal limits, and laboratory findings did not reveal any abnormality.

Contrast-enhanced cranial MRI showed an approximately 4-cm aneurysmal vascular malformation at the left frontal region. The lesion had cortical-subcortical localization, and caused a contra-lateral shift effect, suppression to components of the adjacent lateral ventricles and corpus callosum from the superior, including a surrounding hypointense area in the shape of a rim due to hemosiderin or calcification, and showed flow void signal changes due to prominent fast flow. Drainage veins towards the adjacent dural surfaces could be distinguished within the lesion. MRA and MRV were repeated and revealed that the lesion was supplied with blood from the distal ends of the middle cerebral artery and that drainage veins extended to the superior sagittal sinus, sigmoid sinus on the left, and vein of Galen. DSA evaluation showed that the anterosuperior

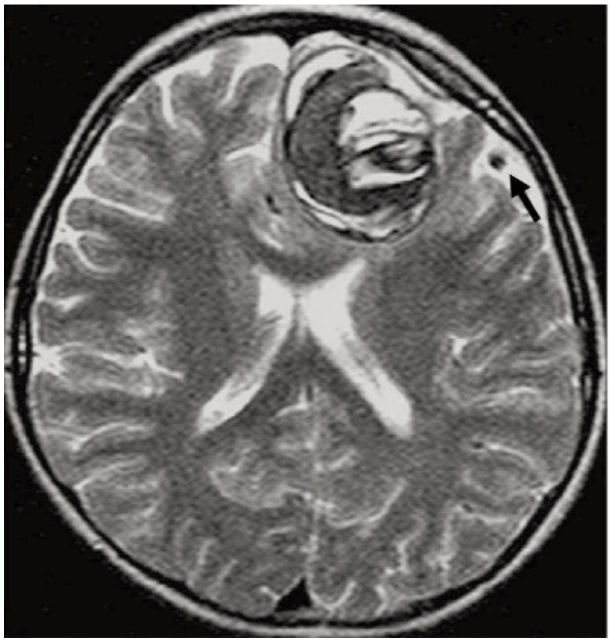

Figure 1A. Axial T2-weighted MRI shows an aneurysmal vascular malformation in the left frontal region surrounded by a hypointense rim and containing signal void areas related to the significantly high flow rate. Dilated drainage veins are observed in the adjacent dural surfaces (arrow).

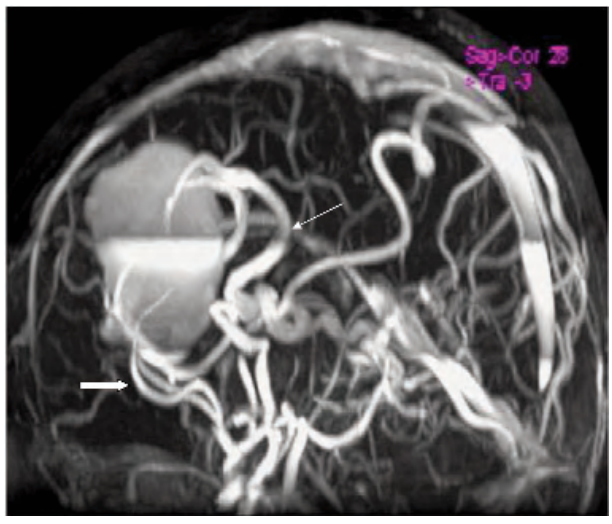

Figure 1B. MRA shows the aneurysmal sac supplied from distal branches of the middle cerebral arteries (thick arrow), and draining veins (thin arrow) extending to the superior sagittal sinus and in the left to sigmoid sinus.

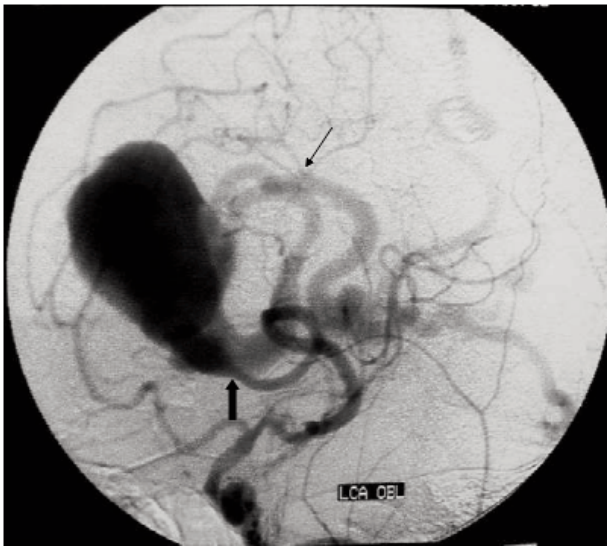

Figure 2A. Angiogram shows the oblique view of the left common carotid artery and a dilated anterior branch of the left middle cerebral artery (thick arrow) opening to a large aneurysmal sac. Drainage veins are also seen (thin arrow).

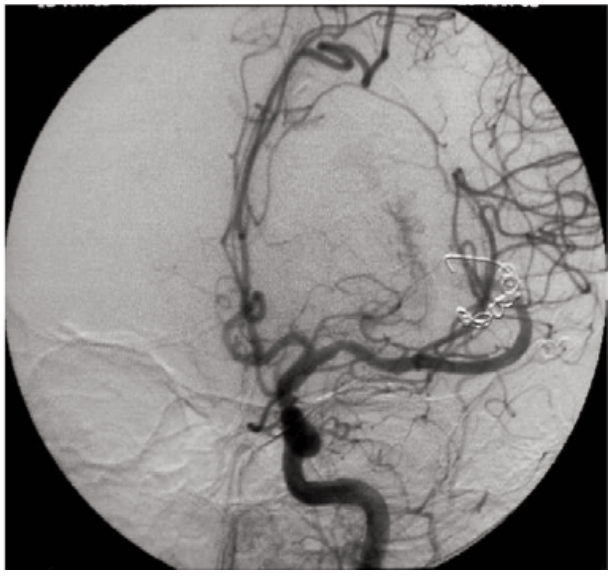

Figure 2B. Anteroposterior view of the left internal carotid artery angiogram shows total occlusion of the distal part of the left middle cerebral artery following embolization. The aneurysmal sac and aberrant veins showed no contrast enhancement.

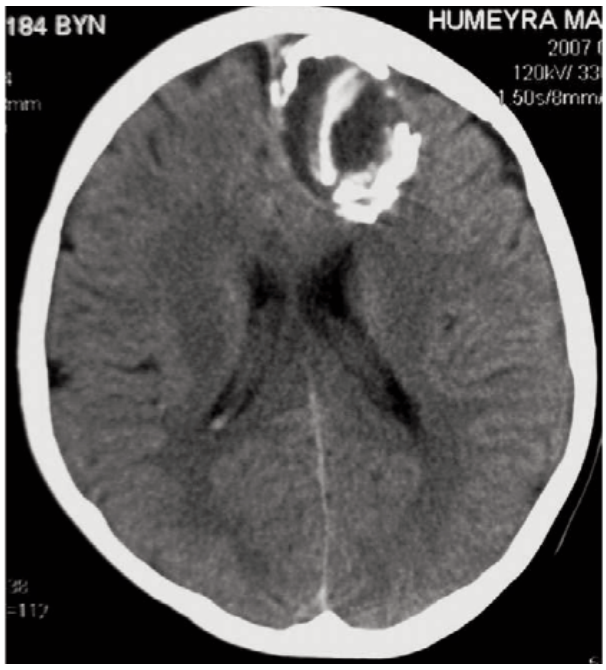

Figure 3A. Follow-up un-enhanced CT shows a slight decrease in the dimensions of the lesion in left frontal region, with increased calcification in the surrounding region.

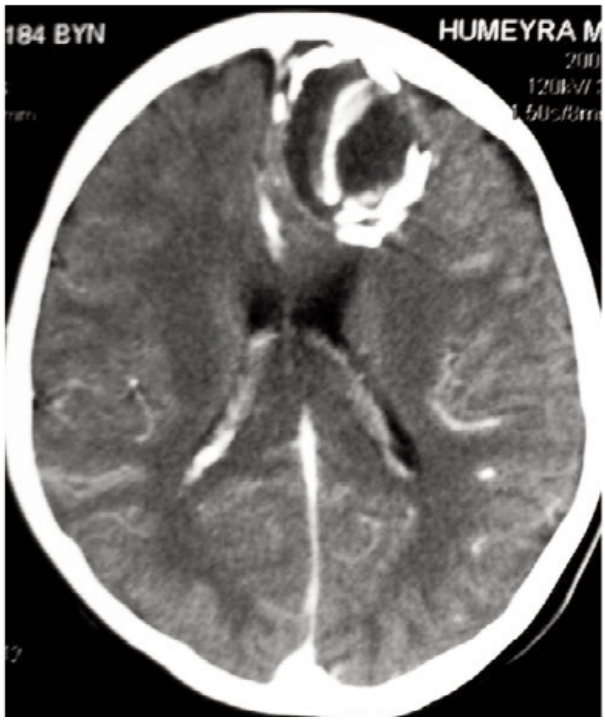

Figure 3B. Contrast-enhanced CT shows no contrast enhancement inside the lesion.

branch of the left middle cerebral artery was dilated and flowed to a large aneurysmal sac in the frontal region and then via the vein of Trolard, and drained into the left sigmoid sinus and superior sagittal sinus. In light of these findings the lesion was considered an AVF and an aneurysmal sac that was approximately 3-4 cm at the localization of the fistula.

Following diagnostic imaging, we decided to occlude the arterial portion of the fistula near the aneurysmal sac. Initially, a guiding catheter was placed into the left internal carotid artery and a microcatheter with 2 markers was forwarded through the guiding catheter into the left middle cerebral artery. After placing the microcatheter just below the fistula, a 5 × 20-cm GDC-18 helical coil was initially placed within the artery. Subsequently, a 3 × 8-cm GDC-18 helical coil was deployed and detached. A follow-up angiogram showed that there was no leakage through the fistula into the aneurismal sac. The patient was then transferred to the intensive care unit. Following embolization, the patient was followed-up for 1 week. During this period anti-edema and antiepileptic therapy was administered to the patient. She did not have any neurological sequelae.

The patient was monitored periodically after treatment. The final CT evaluation revealed that the lesion had decreased in size and that the surrounding calcification had increased. This appearance was accepted as calcification because of its circumferential characteristic without contrast-enhancement and its presence in the next BT examination 1 year later. There was no passage of contrast material into the fistula or venous aneurysmal sac. The patient was followed-up for 2.5 years after the embolization during which time she was asymptomatic.

Discussion

Unlike cerebral AVMs, cerebral AVFs are rare and characterized by a sudden passage between the afferent artery and drainage veins, without a plexiform nidus, and consist of widened vascular structures (1,3). They may be congenital or acquired (3). About 15%-23% of all AVMs are seen in children (1,5) and 4.7% of all intracranial malformations are due to AVFs (1,6). AVFs are rare pathologies; hence, the prevalence of AVFs in children is not well known (1). They may be seen alone; however, they are relatively more prevalent in children with cerebral AVMs (1,3).

AVFs are most commonly seen in the frontal and temporal lobes, and less frequently in the parietal lobes. In supratentorial lesions, the most common afferent artery involved is the middle cerebral artery, followed by the posterior cerebral artery and the anterior cerebral artery (1). They may be single or multiple (3). In multifocal lesions there are different drainage veins at each fistula point, and they are frequently seen together with a giant venous sac (1). In most cases, AVFs drain into the transverse and sigmoid sinuses, and sometimes drain into the vein of Galen (3,4). They are rarely asymptomatic and may show clinical features like congestive heart failure, macrocrania, intracranial hemorrhage, seizure, headache, and focal neurological deficits consistent with the age of diagnosis (1,3). Weon et al. reported that the symptoms and findings that most frequently lead to diagnosis include cardiac failure, epilepsy, and macrocrania. Diagnosis is most frequently made at the age of 5 years (1).

Supportive imaging methods include computed tomography (CT), MRI, and digital subtraction angiography (DSA). CT findings of AVFs are dilated arteries and veins, and a widened sac in the absence of a prominent nidus. There is calcification in the shape of an eggshell at the thrombotic region of the vascular sac or the wall in more than 50% of cases (3,7). Widened vascular structures, dural sinuses into which veins drain, flow characteristics in vascular structures, and the effects of the lesion on surrounding tissues may be evaluated with MRI. Cerebral DSA affords very good visualization of the vascular structures of AVFs and also makes invasive neuroradiological procedures possible (3).

The prognosis of AVFs is poor following conservative treatment; thus, rapid control of the lesions is the goal of treatment, even though they are asymptomatic (1,8). During treatment the fistulas or afferent vascular structures should be completely

obliterated. Successful obliteration may be achieved with surgery or endovascular techniques. Successful surgical treatment has been reported in neonates and infants with symptomatic nongalenic AVFs (1,3,9,10); however, surgical ligation of the fistula is sometimes impossible because widened drainage veins can obscure the fistula. On the other hand, long-lasting shunts may lead to thickening of arteries and drainage veins, complicating definitive evaluation of the fistula area (3). Recent technological advancements and increased neuroradiological expertise have demonstrated the importance of the endovascular approach to the treatment of intracranial AVFs, together with associated giant varices (1,3). Glue (NBCA, cyanoacrylate) is the preferred embolic agent; however, coils are also used (3). Treatment of cerebral AVFs with the transarterial endovascular method decreases both the need for major surgical interventions and the incidence of post-operative hemorrhage, consequently lowering morbidity and mortality (1,3). Care should be taken when using cyanoacrylate so as to prevent the embolizing agent from escaping into the vein and occluding the drainage vein, especially in patients with a single drainage vein.

In the presented case there was a wide single fistula between a dilated superior branch of the left middle cerebral artery and an abnormal dilated drainage vein, together with a large aneurysmal sac at the localization of the fistula. Venous drainage was towards the left sigmoid and superior sagittal sinuses. The afferent artery was covered with 2 coils of different dimensions via microguide and microcatheter. After the procedure, the distal part of the afferent artery was completely occluded, and the aneurysmal sac and drainage veins were not filled. There was no complication during or after the procedure.

In conclusion, cerebral AVFs are rarely encountered vascular lesions in children and may be successfully treated with endovascular treatment. Morbidity and mortality rates associated with endovascular procedures are low.

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