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SPONTANEOUS DISAPPEARANCE OF A CAUDATE NUCLEUS ARTERIOVENOUS MALFORMATION FOLLOWING EXTERNAL VENTRICULAR DRAINAGE: A CASE REPORT AND LITERATURE REVIEW

EKSTERNAL VENTRİKÜLER DRENAJ SONRASI SPONTAN KAYBOLAN KAUDAT ÇEKİRDEK ARTERİOVENÖZ MALFORMASYONU: OLGU SUNUMU VE LİTERATÜR İNCELEMESİ

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ABSTRACT

Brain arteriovenous malformations (AVMs) are congenital vascular anomalies that can cause life-threatening haemorrhages. Spontaneous AVM thrombosis is extremely rare and is not fully understood. A 15-year-old female presented with severe headache and confusion. Computed tomography (CT) revealed intraventricular haemorrhage, leading to urgent external ventricular drainage (EVD). Digital subtraction angiography (DSA) confirmed an AVM located in the right caudate nucleus head, supplied by the lateral lenticulostriate artery and draining into the deep venous system. At the 2-month follow-up, DSA showed complete disappearance of the AVM. This case highlights the rare phenomenon of spontaneous AVM thrombosis, potentially influenced by haemorrhage, venous outflow obstruction, and EVD placement. While spontaneous resolution is possible, long-term imaging follow-up is essential due to the risk of delayed recanalization.

Keywords: Arteriovenous malformation, external ventricular drain, intraventricular haemorrhage, spontaneous thrombosis

ÖZET

Beyin arteriovenöz malformasyonları (AVM), yaşamı tehdit eden kanamalara yol açabilen konjenital vasküler anomalilerdir. AVM'lerin kendiliğinden tromboze olması son derece nadir görülen ve yeterince anlaşılamayan bir durumdur. On beş yaşında bir kız hasta, şiddetli baş ağrısı ve bilinç bulanıklığı ile acil servise başvurdu. Bilgisayarlı tomografi (BT), intraventriküler kanama gösterdi ve acil eksternal ventriküler drenaj (EVD) uygulandı. Dijital substraksiyon anjiyografi (DSA), sağ kaudat çekirdek başına yerleşmiş, lateral lentikülostriat arter tarafından beslenen ve derin venöz sisteme drene olan bir AVM saptadı. İki ay sonra yapılan kontrol DSA'da AVM'nin tamamen kaybolduğu gözlendi ve kendiliğinden tromboz geliştiği düşünüldü. Bu olgu, kanama, venöz drenaj tıkanıklığı ve EVD uygulamasının AVM trombozunu nasıl tetikleyebileceğini gösteren nadir bir örnektir. AVM'lerin kendiliğinden kaybolması mümkün olmakla birlikte, geç rekürrens riski nedeniyle uzun süreli görüntüleme takibi gereklidir.

Anahtar kelimeler: Arteriovenöz malformasyon, eksternal ventriküler drenaj, intraventriküler kanama, spontan tromboz

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INTRODUCTION

Brain arteriovenous malformations (AVMs) are congenital vascular anomalies characterised by abnormal tangles of arteries and veins lacking an intervening capillary network (1). Rupture of AVMs is a well-known cause of intracerebral haemorrhage (ICH) and is the most common cause of ICH in the paediatric population. Symptoms of AVMs, including headache, seizures, and motor weakness, are non-specific and can mimic other neurological conditions (2).

AVMs are classified into five grades based on three features: size (<3 cm, 3–6 cm, >6 cm), location (eloquent vs. non-eloquent), and venous drainage (superficial vs. deep). Diagnosis is typically achieved through computed tomography angiography (CTA), magnetic resonance angiography (MRA), and digital subtraction angiography (DSA), with DSA being the gold standard (3).

In this study, we report a case of an AVM with a small nidus located in the caudate nucleus head, presenting with an intraventricular haematoma that disappeared spontaneously following external ventricular drain (EVD) placement.

CASE PRESENTATION

A 15-year-old woman with no known comorbidities presented to the paediatric emergency unit with headache and confusion. Neurological examination revealed spontaneous breathing, spontaneous eye opening, isochoric pupils with positive light reflex, uncooperativeness, flexor response to painful stimuli, and nonsensical vocalisation (GCS: 8, E2M4V2). The patient was promptly intubated and stabilised.

A non-contrast cranial computed tomography (CT) scan demonstrated diffuse intraventricular haemorrhage. An EVD was immediately placed, with the evacuation of the high-pressure haemorrhagic fluid. The drainage rate was adjusted to 10–15 cc/h. A contrast-enhanced cranial CT angiography scan was subsequently performed, revealing an AVM in close proximity to the right caudate nucleus head and near the EVD catheter (Figure 1).

ostic digital subtraction angiography (DSA) confirmed an AVM in the same region, receiving arterial supply from the lateral lenticulostriate artery. The nidus measured approximately 6 mm in diameter, with a 1.2 mm aneurysm within it. Venous drainage occurred via the deep circulation through the thalamostriate vein. The AVM was classified as Spetzler-Martin Grade 2 and Lawton-Young Score 1.

The patient was transferred to the intensive care unit (ICU), where sedation was gradually reduced. By the second day post-haemorrhage, she was extubated. The patient's neurological examination was intact (GCS: 15),

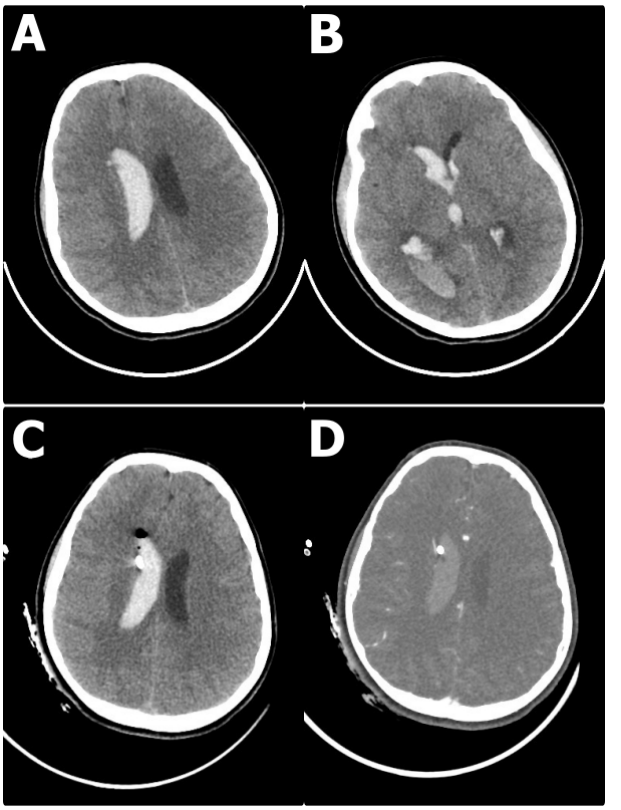


Figure 1: (A, B) Preoperative CT scans demonstrating acute intraventricular haemorrhage. (C) Postoperative CT scan confirming persistent intraventricular haemorrhage with the EVD catheter positioned within the ventricle. (D) Postoperative CT angiography revealing intraventricular haemorrhage, a suspected AVM nidus at the head of the right caudate nucleus, and the close proximity of the EVD catheter to the AVM drainage vein within the ventricle.

with no motor deficits or cognitive impairment.

Follow-up imaging showed a gradual reduction in intraventricular haematoma volume. The EVD was removed on the seventh day post-haemorrhage. The patient had an uneventful recovery, with no wound infections, fever, or additional complaints. She was discharged on day 20 with a scheduled follow-up DSA after 2 months.

At the 2-month follow-up, repeat DSA revealed complete disappearance of the previously identified AVM adjacent to the right caudate nucleus head (Figure 2).

DISCUSSION

Management options for AVMs include microsurgical resection, stereotactic radiosurgery, endovascular embolisation, or a combination of these approaches. The Spetzler-Martin and Lawton-Young grading systems guide treatment selection by assessing the AVM size, location, venous drainage, patient age, and haemorrhage history (4).

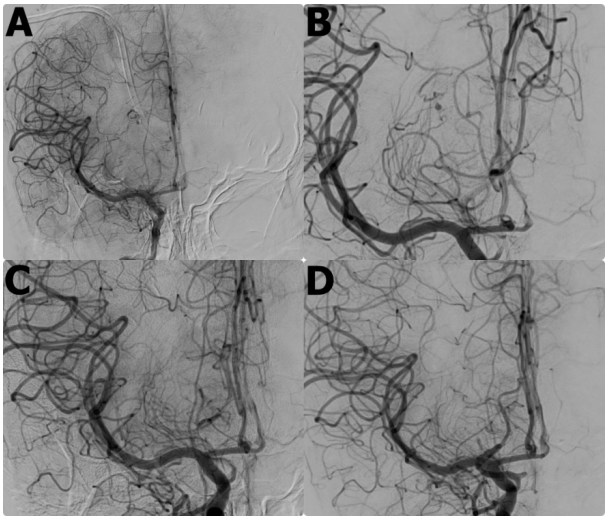


Figure 2: (A, B) Preoperative diagnostic DSA scans showing an AVM adjacent to the right caudate nucleus head, supplied by the lateral lenticulostriate artery. The nidus measured approximately 6 mm in diameter, with a 1.2 mm aneurysm identified within it, and drainage occurred via the thalamostriate vein. The AVM was also noted to be in close proximity to the EVD catheter and its drainage vein within the ventricle. (C, D) Follow-up DSA at 2 months demonstrating complete disappearance of the previously identified AVM near the right caudate nucleus head.

In the Spetzler-Martin grading system, microsurgical resection is typically preferred for low-grade AVMs (Grades I–III) because of the relatively low surgical risk. Conversely, high-grade AVMs (Grades IV–V) may be unsuitable for surgery, and alternative treatments such as stereotactic radiosurgery or endovascular embolisation are preferred (5). Lawton et al. introduced the Supplemented Spetzler-Martin grade, which incorporates additional factors such as patient age, haemorrhage history, and nidus type to improve surgical risk assessment (4).

Spontaneous AVM disappearance

Spontaneous regression of brain AVMs is exceptionally rare, with an estimated incidence of 0.1%. Small AVMs with a single superficial draining vein and prior haemorrhage have a higher likelihood of spontaneous thrombosis (6).

Following an initial haemorrhage, the mass effect and associated hemodynamic changes may lead to reduced blood flow to the AVM. The subsequent gliotic reaction altered venous drainage and promoted thrombosis. In AVMs with a single draining vein, the lack of alternative outflow pathways may increase the susceptibility to complete thrombosis. This process is believed to be gradual, with thrombosis developing over time rather than as a sudden event (7).

In this case, the presence of a single deep draining vein and the history of previous haemorrhage likely contributed to the spontaneous AVM thrombosis. Additionally, the proximity of the EVD catheter to the AVM nidus may have facilitated thrombosis, potentially due to localised hemodynamic alterations or inflammatory responses.

Clinical implications and follow-up

The annual haemorrhage rate of AVMs is estimated to be 2.2%–4.5% per 100 patient-years, increasing significantly after an initial rupture. The risk of rehemorrhage within the first year post-rupture ranges from 6% to 15.8%, emphasising the importance of early intervention in high-risk cases (8). However, given the extremely low likelihood of spontaneous AVM obliteration, conservative management should not be considered a primary strategy. Patients who opt for non-operative management should be informed of the rare but possible outcome of spontaneous resolution (9).

Long-term follow-up is essential because cases of delayed AVM recanalization have been reported up to 39 months after the initial disappearance (10). In this case, no recanalization was observed during the six-month follow-up period. However, continued surveillance with magnetic resonance angiography (MRA) has been recommended to rule out late recanalization.

CONCLUSION

The spontaneous disappearance of brain AVMs is an extremely rare phenomenon, likely influenced by a combination of haemorrhage-induced hemodynamic changes, venous outflow obstruction, and gliotic responses. In this case, the presence of a single deep draining vein, prior bleeding, and the proximity of the EVD catheter to the AVM nidus may have contributed to the thrombosis. While spontaneous resolution is possible, it should not be considered a primary management strategy due to the unpredictable risk of rebleeding or delayed recanalization. Long-term radiological follow-up remains crucial to ensure that the AVM does not reappear, and further research is needed to better understand the mechanisms behind spontaneous AVM obliteration.

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