



## Case Report

# Pediatric intracranial calcified arteriovenous malformation: A case report

Fatima Mustansir<sup>1</sup>, Meher Angez<sup>1</sup>, Mohammad Hamza Bajwa<sup>2</sup>, Saira Fatima<sup>3</sup>, Syed Ather Enam<sup>2</sup>

<sup>1</sup>Medical College, Aga Khan University, <sup>2</sup>Department of Neurological Surgery, Aga Khan University, <sup>3</sup>Department of Histopathology, Aga Khan University, Karachi, Pakistan.

E-mail: Fatima Mustansir - fatimamustansir5@gmail.com; Meher Angez - meher.angez@scholar.aku.edu; Mohammad Hamza Bajwa - mhamzabajwa95@gmail.com; Saira Fatima - saira.fatima@aku.edu; \*Syed Ather Enam - ather.enam@aku.edu



### \*Corresponding author:

Syed Ather Enam,  
Department of Neurological  
Surgery, The Aga Khan  
University Hospital, Karachi,  
Pakistan.

fatimamustansir5@gmail.com

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## ABSTRACT

**Background:** Brain arteriovenous malformations (AVMs) are intracranial lesions that consist of a complex tangle of abnormal blood vessels. They can occasionally become hard and calcified. This may render these lesions difficult to resect and lead to neurological complications. There are very few reported cases of calcified brain AVMs in the literature.

**Case Description:** We report the case of an 11-year-old patient who presented with headaches and seizures exacerbated in the past 3 months. Preoperative imaging confirmed a large, right parasagittal AVM, with significant internal calcifications seen on the computed tomography angiogram. We performed a successful microsurgical resection of the calcified AVM and confirmed the diagnosis on histopathological analysis.

**Conclusion:** Dense internal calcifications within AVMs are a clinical rarity and can be challenging cases for microsurgical resection.

**Keywords:** Aneurysm, Arteriovenous malformation, Calcified arteriovenous malformation, Cerebrovascular

## INTRODUCTION

Brain arteriovenous malformations (AVMs) are rare intracranial vascular lesions consisting of a nidus of vessels traversed by arteries and veins without intervening capillaries. This anatomical arrangement leads to extensive shunting of blood between the arterial and venous systems.<sup>[1,2]</sup> The incidence of AVMs is around 1/100,000 people annually.<sup>[1]</sup> Approximately 12–18% of AVMs are found in children.<sup>[4,10]</sup> Despite occurring less frequently in the pediatric population, ruptured AVMs represent a serious risk as they are commonly implicated in spontaneous intracerebral hemorrhage (ICH) in children.<sup>[14]</sup>

Dystrophic calcifications within AVMs are a common finding on radiographs, particularly on computed tomography (CT) scans, as they have an increased sensitivity for calcifications.<sup>[3]</sup> These calcifications are usually faint and appear as lacelike punctate densities or curvilinear streaks.<sup>[2,17]</sup> However, large solitary or multiple intracranial calcifications, also referred to as “brain stones” or “cerebral calculi,” are less frequently encountered.<sup>[8]</sup> To the best of our knowledge, few reports of brain stones exist within the literature,<sup>[7,8,17]</sup> resulting in uncertainty regarding treatment

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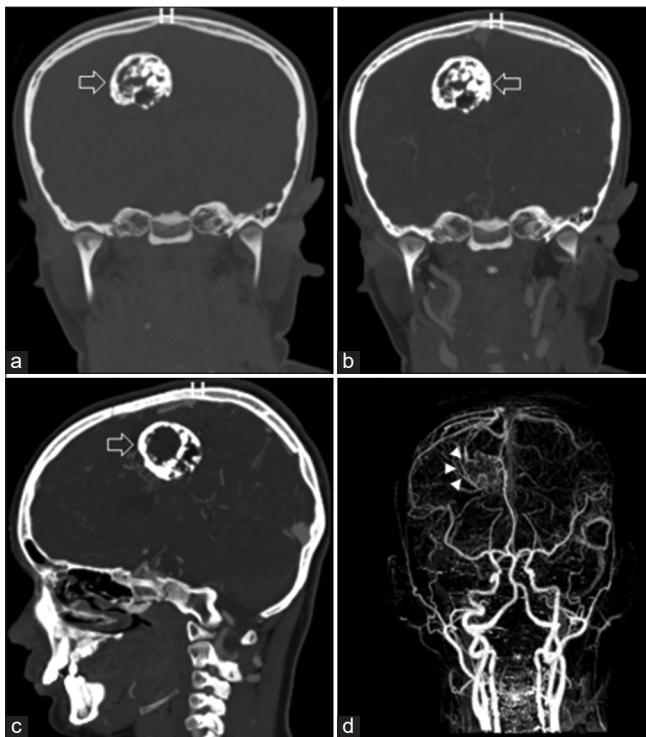
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strategies for calcified AVMs. In this report, we present a case of a pediatric patient who was found to have a calcified AVM which was successfully resected.

## CASE PRESENTATION

An 11-year-old boy presented to the outpatient neurosurgery clinic at a tertiary care hospital with a 3-month history of seizures and severe headaches. The patient's family described two episodes of generalized tonic-clonic seizures lasting for 2–3 minutes each. The patient had also reported experiencing intermittent headaches for the past 2 years which had gradually increased in severity over the past 3 months. These more recent headaches were dull in character, radiating to the neck, and associated with nausea and vomiting. He had a history of asthma that was controlled on medications. He was born to consanguineously married parents. The patient's family history was otherwise unremarkable. On initial presentation, his physical examination, including a complete neurological examination, was unremarkable.

Initial magnetic resonance imaging (MRI) from an outside hospital showed a right parasagittal lesion with postcontrast

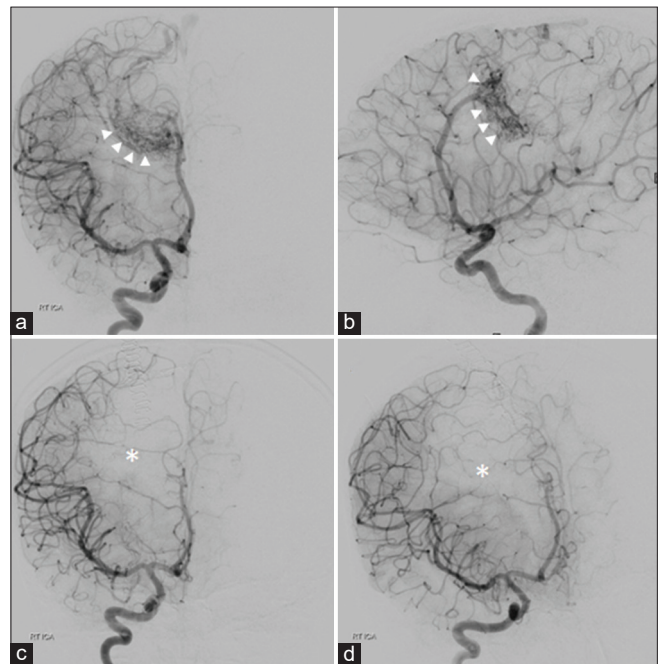


**Figure 1:** Computed tomography head with coronal noncontrast (a), coronal postcontrast (b), and sagittal postcontrast (c) images showing a well-defined heterogeneous lesion in the right parasagittal region (white arrows) with significant internal calcifications. Flow voids are suggestive of an arteriovenous malformation (AVM) with the nidus measuring approximately  $3.8 \times 3.4 \times 3.7$  cm. CTA with coronal reconstruction (d) confirms the location of the AVM in the right frontal parasagittal region (white arrowheads).

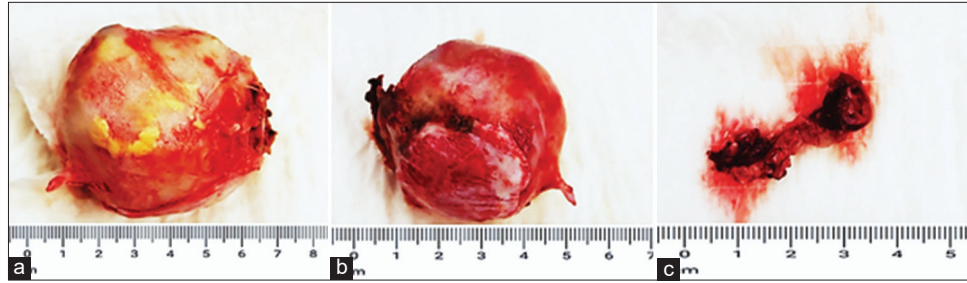
enhancement that was suggestive of a highly vascular pathology. Differential diagnoses based on radiology included hemangiopericytoma and a lipomatous lesion. He was prescribed divalproex sodium, which was unsuccessful in controlling his seizures.

Based on these findings, the patient underwent a CT angiogram (CTA) for further investigation of a vascular etiology. CTA revealed a well-defined heterogeneous lesion in the right parasagittal region with significant internal calcification [Figure 1]. Flow voids were suggestive of an AVM. A digital subtraction angiogram (DSA) confirmed the diagnosis of a large, calcified AVM in the right parasagittal location, with the nidus located at its anterolateral margin. This was supplied by the right anterior cerebral artery, mainly through branches of the right pericallosal artery [Figures 2, 2a and b]. There were no feeders arising from external carotid arteries, left internal carotid artery, or posterior circulation. The AVM was seen to be draining into the superior sagittal sinus through a superficial cortical draining vein.

After discussing the risks and benefits of stereotactic radiosurgery and conventional surgical resection, the patient's family opted for surgery. The patient underwent a neuronavigation-guided right frontal craniotomy under



**Figure 2:** Anteroposterior (a) and lateral (b) right internal carotid angiogram showing a lesion (white arrowheads) supplied by the distal branches of the right cerebral artery, mainly branches of the right pericallosal artery. The arteriovenous malformation (AVM) is shown to be draining into the superior sagittal sinus through a superficial draining vein. Postoperative angiography (c and d) shows complete resection with no arterial blush seen at the prior site of AVM (white asterisk).



**Figure 3:** Gross specimens of the calcified arteriovenous malformation (a and b) and its nidus (c) after surgical resection.

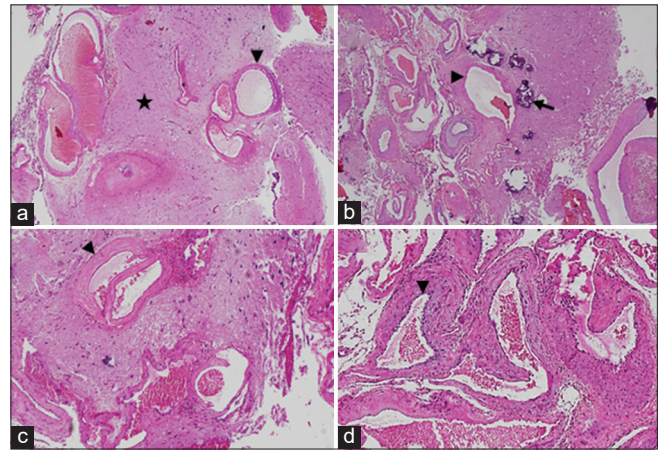
general anesthesia. Intraoperative findings included a hard, calcified, and well-encapsulated lesion which was adherent to the falx cerebri. The nidus of the AVM was confirmed to be at the anterolateral margin. The lesion was separated from the brain parenchyma along the falx cerebri and collateral vessels were coagulated. The AVM was resected *en bloc* along with its nidus [Figure 3]. Histopathology confirmed a calcified AVM with no evidence of malignancy [Figures 4 and 5]. The patient lost approximately 500 ml of blood but there were no intraoperative complications.

The patient remained stable postoperatively and was subsequently shifted to the intensive care unit for overnight monitoring. On the 1<sup>st</sup> postoperative day, he noticed difficulty in moving his left arm and leg against gravity. Power in his left upper extremity was 3/5 while power in his left lower extremity was 2/5. He did not have any additional neurological deficits. He was managed with intravenous fluids, antibiotics, anti-emetics, analgesics, and steroids. In addition, he received chest and limb physiotherapy and occupational therapy. A DSA on the 2<sup>nd</sup> postoperative day showed no evidence of residual disease or arterial blush [Figures 2c and d]. The patient's left-sided weakness continued to improve and he was discharged from hospital on the 4<sup>th</sup> postoperative day. He continued to receive physiotherapy. On his clinic follow-up after 2 weeks, the patient reported improving motor function with resolution of his previous symptoms.

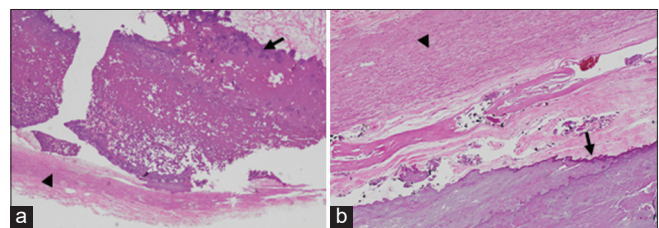
## DISCUSSION

While intracranial calcifications are not an uncommon finding in a variety of pathologies, what constitutes a “brain stone” or “cerebral calculus,” especially in terms of size and morphology, remains to be clearly determined. Nearly 60 years ago, Tiberin and Beller<sup>[16]</sup> defined brain stones or cerebral calculi as “large, solitary or multiple, well-circumscribed, bony hard areas of pathologic intracerebral calcification” representing the end stage in the evolution of certain nonneoplastic space-occupying lesions.

Brain stones can be classified based on their location, either extra-axial or intra-axial, and etiology. While extra-axial calcified lesions are usually tumors or exaggerated



**Figure 4:** Photomicrographs of specimen sent: (a and b) H&E (×4) images showing normal glial tissue (black star) with malformed blood vessels. Calcified deposits are seen within the vessels. (c and d) H&E (×10) images showing abnormally dilated vessels with smooth muscle in the walls. Blood vessels are highlighted with black arrowheads. Similarly, black arrows indicate calcified deposits.



**Figure 5:** (a) H&E (×4) and (b) H&E (×10) images showing completely calcified abnormal vessels. Blood vessels are highlighted with black arrowheads. Similarly, black arrows indicate calcified deposits.

physiological calcifications, intra-axial lesions include vascular, neoplastic, congenital, infectious, endocrine, or metabolic anomalies.<sup>[5,8]</sup> In addition, Gezeran *et al.*<sup>[8]</sup> suggested that lesions >1 cm be classified as brain stones while lesions smaller than 1 cm be classified as calcifications, as all six cases of brain stones in their series were >1 cm. AVMs are the second most frequently calcified lesion, after cavernous malformations.<sup>[8]</sup> These calcifications are either found along the serpentine vessels

of the AVM, in the adjacent brain parenchyma, or within the nidus.<sup>[5]</sup>

AVMs can lead to a variety of symptoms such as hemorrhagic stroke, seizures, headaches, and focal neurologic deficits. Asymptomatic AVMs may also be detected incidentally as noninvasive imaging modalities become more readily available and advanced.<sup>[11]</sup> The same holds true for brain stones – they may present with seizures or be discovered incidentally on imaging.<sup>[8]</sup> Our patient also presented with seizures and headaches.

Cerebral angiography, using subtraction and magnification, remains the gold standard investigation for diagnosing AVMs. A CT scan can be used to initially image the AVM to detect ICH and calcifications. CTA provides further details about the location, size, and drainage of the AVM.<sup>[15]</sup> In addition, MRI has a similar sensitivity to CTA in detecting AVMs<sup>[9]</sup> and has been shown to have a low false-negative rate.<sup>[13]</sup> However, in our case, MRI was unable to detect the presence of an AVM and was instead suggestive of a neoplastic lesion, such as a hemangiopericytoma or a lipomatous lesion. The diagnosis of a calcified AVM became apparent on CTA and DSA.

At present, available treatment options for brain AVMs include microsurgical resection, stereotactic radiosurgery, and endovascular embolization, either performed as primary therapy or as part of a multimodal treatment plan. The goal of treatment is complete elimination of the nidus and arteriovenous shunt, as partial nidal obliteration carries a substantial risk of hemorrhage.<sup>[6]</sup> Our patient underwent microsurgical resection of the calcified lesion and AVM as the primary treatment. Calcifications in and around the AVM rendered it more difficult to resect. Florian *et al.*<sup>[7]</sup> reported of a large calcified brain AVM shares certain similarities with our case. Although their patient was a 55-year-old adult, he also reported seizures and headaches as the manifestation of a large, densely calcified AVM. The calcified lesion was readily visible on preoperative CT scan and CTA. The patient's lesion was completely resected through right temporoparietal craniotomy, in what the authors describe as a challenging surgery due to problems achieving hemostasis because of the rigid and calcified vessels.

This was our first experience of managing a patient with a calcified AVM. While the calcified nature of the lesion did not present a substantial increase in the difficulty of resection, we were faced with a diagnostic conundrum when the AVM was initially diagnosed as a neoplastic lesion. We were able to resect the lesion in its entirety, including the nidus, leading to improvement in the patient's symptoms.

## CONCLUSION

Large calcified intracranial lesions or brain stones are a rarely encountered pathology. Brain stones in association with various intracranial lesions have been seldom reported

in the literature and even more infrequently in association with AVMs. We report the case of a pediatric patient with a large densely calcified AVM, who underwent successful microsurgical resection of the lesion.

## Acknowledgments

Nil.

## Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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## Conflicts of interest

There are no conflicts of interest.

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