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Case Report

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Surgical Management of a Thalamic Arteriovenous Malformation in a Young Male in a Low Resource Setting: An Illustrative Case Report

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ABSTRACT

Introduction: The natural history of patients with arteriovenous malformations (AVMs) suggests that 10-17% die due to hemorrhage and 40-50% experience serious physical deterioration of their working capacity in their 20-40-year period after presentation and treatment. Deep basal ganglia and thalamus lesions account for 3-12% of all AVMs and some literature suggests that these lesions have a more hostile clinical course.

Case Presentation: 16-year-old male, a student with no known chronic illnesses came to our hospital as a referral from Lubaga hospital. He presented with fifteen days' history of sudden onset of severe headache with no known relieving or exacerbating factors. This was associated with episodes of vomiting, altered mentation, right sided weakness, however there was no history of convulsions, blurring of vision or trauma. This was the first time for this kind of presentation. On admission: He was lethargic, moderately dehydrated, not pale nor jaundiced. BP was 109/52mmHg, HR of 60bpm, SPO2=96% on room air. Glasgow Coma Scale score of 14/15, V=4, PEAL, soft neck, normal muscle bulky with a power of 2/5 on the left side and 4/5 on the right side. Successful surgical resection of the AVM was done at Mbarara regional referral Hospital-Uganda. Currently the patient is back in the community and has returned to school.

Conclusion: Children who present with intracranial hemorrhage from a previously undiagnosed AVM have a 12% chance of sudden death. Clinical triggers of hemorrhage are unpredictable, and all fatal cases were in locations with high risk of potential herniation. These data support a proactive, aggressive approach toward definitive treatment of AVMs in children and adults. Patients with diagnosed AVM can once again get a chance to survive like other children in the community in LMICs like Uganda by undergoing open surgery, although advanced opportunities are still limited.

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Introduction

Cerebral arteriovenous malformations (AVMs) are among the major causes of spontaneous intracerebral hemorrhage. AVMs in adult's population, account for 1.4% of all adult intracranial hemorrhages, they also represent the cause in 20–50% of spontaneous intracranial hemorrhages in children [1]. Globally, the prevalence of cerebral AVMs is less than 1%, most of these cases are congenital, only 3–19% of those treated for AVMs are children [2]. Arteriovenous malformations are the commonest cause of intracerebral hemorrhage in children. Different management options exist including: open microsurgery, endovascular embolization, stereotactic radiosurgery, or a combination of these treatments [3]. The aim of all these therapies is to eliminate the nidus of AVM, thus removing the risk of future hemorrhage.

The natural history of AVMs has not been fully determined especially for children. Studies suggest that the annual risk of hemorrhage in children is higher than that of adults (2–4% vs. 1–3% per year) [4]. Each hemorrhagic event carries a 5–10% risk of mortality and a morbidity of up to 50% [5]. Pediatric AVMs have a higher propensity of presenting with hemorrhage than adult AVMs, suggesting there may actually be a higher hemorrhage risk in children [6]. Treatment for deep basal ganglia and thalamic lesions has been limited to non-operation management like stereotactic radiosurgery (SRS) and gamma knife. Anecdotal evidence suggests few cases have been managed surgically with reported good outcomes. We report a case of surgical management of a deep basal ganglia/thalamic lesion management of a 16-year-old male at Mbarara regional referral hospital with good outcomes.

Case Presentation

Patient Description

The patient is a 16-year-old male with no known chronic illnesses and no reported relevant social, environment or family history. He was referred to our hospital from Lubaga hospital in Kampala due to financial constraints. He presented with fifteen days' history of sudden onset of severe headache with no known relieving or exacerbating factors. This was associated with episodes of vomiting, altered mentation and left sided weakness, however, there was no history of convulsions, blurring of vision or trauma. This was the index episode for this kind of presentation.

In the review of other systems, there was nothing remarkable.

On admission: He was lethargic, moderately dehydrated, not pale or jaundiced. BP was 109/52mmHg, HR of 60bpm, SPO2=96% on room air. Glasgow Coma Scale score of 14/15, V=4, PEAL, soft neck, normal muscle bulky with a global power of 2/5 on the left and 4/5 on the right side

Examination of Other Systems: Normal findings
Laboratory tests showed normal full hemogram, all electrolytes were within normal ranges.
And a Cranial MRI done on (29/12/2021) from a facility in Kampala (5-hour drive from our hospital) showed: Right basal ganglia hemorrhage with a midline shift.

Investigations: Imaging:

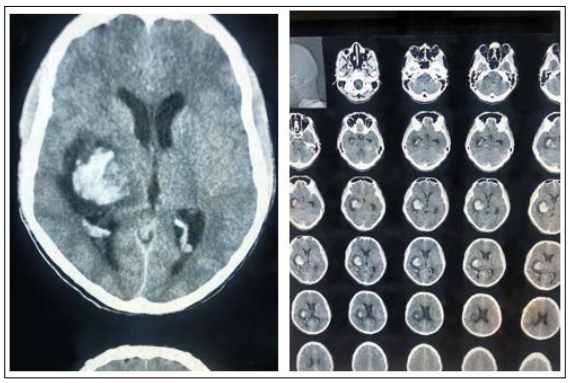

Figure 1: Shows Axial CT scan series

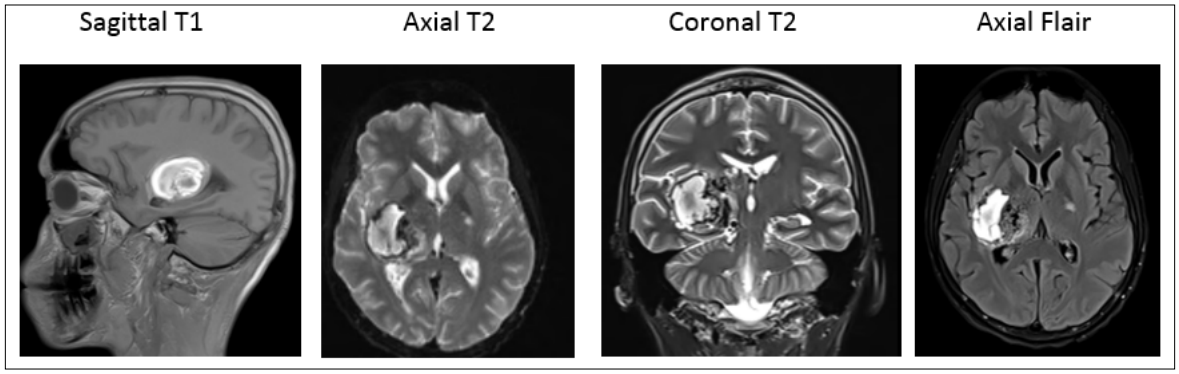

Figure 2: shows Vascular bleed is seen in the right thalamo-capsular region and extending into the right corona radiate. This is seen as hyper intense on T1 and Flair and hypo intense on T2W images. Minimal perilesional edema is seen without any mass effect. Multiple dilated tortuous vessels are seen on the postero-medial aspect of the lesion with prominent internal cerebral veins and straight sinus. Normal ventricular system

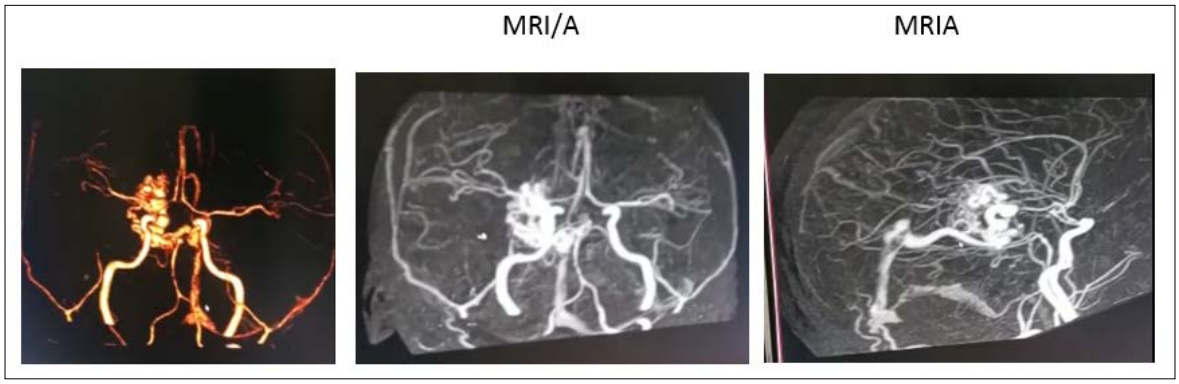

Figure 3: Shows a 3.0cm right temporoparietal AVM with feeding arteries from the 3rd segment of PCA with the Galenic complex, straight and transverse sinuses draining the AVM, there are no associated aneurysms

Diagnosis. Right thalamic AVM-Spetzler-Martin Grade III, Supplementary grading IV

Approach for Surgery

Preparation of the patient for craniotomy and AVM resection was done, pre-anesthetic review done on 10/1/2022 and surgery done on 10/1/2022.

Diagnosis: SM III, Supplementary AVM grade IV

Surgery: Rt transcortical (ITG/MTG)

Incision: Marked a horse shoe incision and infiltrated with local anesthetic and 1:100,000 adrenaline solution.

Reflected myocutaneous flap inferiorly basing onto the temporal floor.
High speed drill, 5x5 temporal craniotomy made.
Dura opened in an inverted U basing on the temporal floor.
Brain moderately tense, identified the STG, MTG and ITG.
Corticotomy made at the posterior MTG and hematoma encountered at 1.5cms below the cortical surface.
Utilized surgical trajectory provided my hematoma to access the AVM nidus.
Feeders from the 3rd segment of the PCA identified and arterialized draining veins to the Galenic complex and onto the straight and right transverse venous sinuses.
Resected AVM starting with the arterial feeders. Left vascular clips onto deep draining veins. Gross total resection achieved.

Post op CT confirmed gross resection and patient improved post op (pre op GCS vs Post op GCS)

Long term follow up- 1year: hemiparesis much resolved and patient back to attending school with minimal fine motor deficits in the left hand

STG=Superior temporal gyrus
ITG=Inferior temporal gyrus
MTG=middle temporal gyrus

Intraoperative Images are Shown in Figure 4.

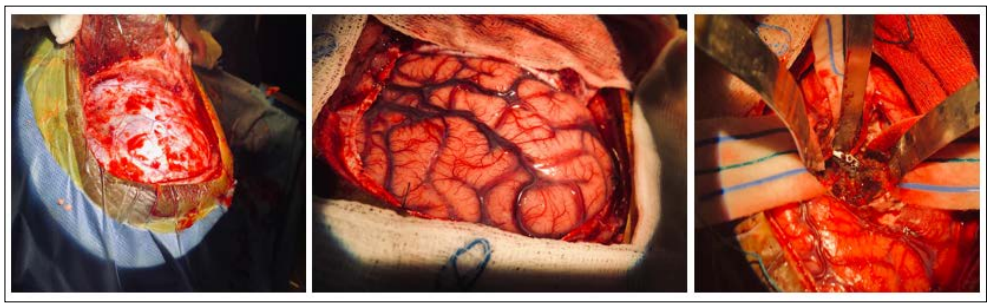

Figure 4: Shows the skin flap made and the clipping of the AVM

The patient spent one night in ICU and the immediate post-operative period was uneventful. He had a GCS=15/15, power in the left limbs was3/5 and on the right side 4/5. Over the course of hospitalization, power greatly improved to 4/5 in all limbs. Six months of follow-up period, the patient had regained full motor function.

He was discharged on post-operative day five. He has recuperated well at home and recently returned to school.

Figure 5: On day 3, postoperative cranial Ct scan, Figure 5, was done and showed; metallic clips are seen in the right temporoparietal area. Overlying cortical sulci are mildly effaced with minimal midline shift to the left. Ventricular system appears normal.

Discussion

Quite a number of literature has been put before us concerning AVMs and it has been noted that the management options of AVMs in children and adults may differ. Children with symptomatic AVMs are at a higher likelihood of bleeding (80%) than adults (50%–65%) [7]. Basal ganglia and thalamic arteriovenous malformations (AVMs) show a poor natural history and have proven difficult to manage especially in LMICs where there is lack of sophisticated treatment options, like embolization [8]. A study done at Boston children's hospital showed that with subsequent radiographic evidence of posterior fossa AVMs, a 57% fatality rate was seen [7].

AVMs initially were thought to be solely congenital lesions that arise during the third gestation week secondary to disordered embryogenesis, where primordial vascular channels fail to differentiate into mature arteries, capillaries, and veins, forming direct arteriovenous shunts without intervening capillary beds [9]. However, reports of the novo formation and recurrence of AVMs after treatment do exist and raise quite a number of questions about the true epidemiology of these vascular lesions [10].

There's a hypothesis that was tested by Hashimoto et al about AVMs being dynamic lesions, he found an approximately sevenfold rise in the number of non-testing endothelial cells in AVMs versus endothelial cells of normal cortical vessels. Additionally, AVM vessels from younger patients showed a trend to increase Ki-67 index, providing some evidence of increased endothelial cell turnover in AVMs [11]. Most AVMs occur sporadically however there are documented familial cases and syndromes associated with vascular malformations. Children with Osler-Weber-Rendu disease have a 7.9% risk for developing symptomatic AVM and potentially multiple AVMs. Wyburn-Mason syndrome may present with a visual pathway or midbrain AVM and ipsilateral facial nevi [3].

Mostly, cerebral AVMs are diagnosed at age 20–40 years, and the most frequent clinical presentation both in adults and children is hemorrhage [3]. However, children present with hemorrhage more commonly than adults [12]. Seizure is the second most common presentation in children accounting for 15% of all pediatric patients' presentation. Children with large vascular malformations may present with symptoms caused by mass effect from a large draining vein or varix [3].

Children who present with intracranial hemorrhage from a previously undiagnosed AVM have a 12% chance of sudden death [7]. Clinical triggers of hemorrhage are unpredictable, but subsequent radiographic evidence of posterior fossa AVMs presented a 57% fatality rate in a study done at Boston Children's Hospital, and all fatal cases were in locations with high risk of potential herniation [7]. These data support a proactive, aggressive approach toward definitive treatment of AVMs in children and adults.

We present a case of thalamic AVM managed successfully at mbarara regional hospital with open surgery and AVM total resection.

Conclusion

We present a 16 y/o male presenting with an acute hemorrhage from a thalamic AVM who was successfully treated at MRRH with craniotomy and AVM resection.

Children with intracranial hemorrhage from a previously undiagnosed AVM have a 12% chance of sudden death, in Uganda it's even higher due to the lack of advanced diagnostics and therapeutic technology (Angiography, endovascular treatment and radiosurgery). Despite that, specifically selected patients with AVM can safely be surgically managed from MRRH-Uganda.

Patients with diagnosed AVM can once again get a chance to survive like any other children in the community in LMICs like Uganda by undergoing open surgery methods though advanced techniques are still limited.

Declarations

Ethics Approval and Consent to Participate

Written informed consent for publication was obtained from the participant's biological mother of this case report

Consent for Publication

Written informed consent for publication of this case report was obtained from the participant's biological mother.

Availability of Data and Materials

Not applicable for this paper

Competing Interests

There are no competing interests for this paper

Funding

This case report was not funded

Authors' Contributions

Ssembatya Joseph Mary. Wrote the manuscript and also participated in surgery.
Blessing Michael Taremwa. Was the main surgeon and also provided the surgical notes, reviewed the manuscript.
Mukuye Simon. Reviewed the manuscript and provided corrections.
Walt Johnson. Reviewed the manuscript and provided corrections.
David Kitya. The head of division of Neurosurgery at Mbarara regional referral hospital.

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