

Spontaneous Ruptured Brain Arteriovenous Malformation in Adolescent Patient: Case Report

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ABSTRACT:

Introduction: Brain arteriovenous malformations (bAVMs) are one of the most common causes of intracranial hemorrhage in the adolescent. AVMs are relatively rare as congenital intracranial abnormalities but these lesions are becoming frequently reported. AVMs generally has no any symptoms and only recognized after intracranial or subarachnoid hemorrhage occurs. This disease usually gives symptoms of headaches and seizures without cause. However, along with the development of medical technology, arteriovenous malformation lesions are common.

Case report: This 24 year old woman in unconscious with right hemiparesis and atypical chronic progressive headache with vas 8-9, without aura. Previously the patient had a seizure with a duration of 5 minutes, rigid all over body. The patient had no prior history of trauma, infection, hypertension, diabetes or stroke. GCS E2M5Vaphasia with motoric aphasia, motoric and sensory deficits in the form of right hemiparesis, paresis of N.VII, increased physiological reflexes, and positive Babinsky reflex. CT-Scan Angiography imaging of the head showed intraparenchymal bleeding in the left temporoparietal lobe with perifocal edema, AVM in the left temporalis region with a nidus with a feeding artery from the left MCA and a draining vein from the left transversesinus. The patient was referred for further vascular intervention.

Discuss: AVMs was previously considered a congenital malformation with risk of bleeding in anomalous tissue or nidus occurring within the brain parenchyma. Rupture of the artery and discharge into the subarachnoid space causes a sudden increase in ICP, cerebral vascular vasospasm resulting in global and focal brain dysfunction. In addition, with the increasing use of non-invasive intracranial imaging, AVMs can be detected directly.

KEYWORDS: brain; arteriovenous malformation (AVM); adolescent; spontaneous;

INTRODUCTION

Arteriovenous Malformation is a congenital abnormality in which the arteries and veinson the surface or parenchyma of the brain are connected directly to each other without going through capillaries. The lesion consists of three components, feeding arteries, nidus and draining vein. Arteriovenous malformations are believed to be abnormalities in vascular development between the fourth and sixth weeks of gestation.¹ Brain arterio-venous malformations are relatively rare intracranial disorders. Lesions that occur as a result of this congenital abnormality appear and are recognized after bleeding occurs.

The incidence of AVMs is approximately 1 per 100,000 per year in the population and the prevalence in adults is approximately 18 per 100,000. AVMs account for between 1 and 2% of all strokes, 3% of young adult strokes, 9% of subarachnoid hemorrhages and 4% of all primary intracerebral hemorrhages.² The exact cause of AVMs is currently unknown. But although their pathophysiology remains unknown, AVMs are generally considered to be congenital lesions that arise from embryological vascular development resulting in structural differences in capillaries and anomalies between arteries and veins.³

AVM is generally a disease that does not show any symptoms and is only discovered after intracranial or subarachnoid hemorrhage occurs. AVM lesions are usually detected by physical examination and then confirmed by CT or MRI. The best diagnostic tool is cerebral angiography. Cerebral angiography is useful for delineating the anatomy of the AVM and determining the characteristics of the blood vessels, and assists the surgeon in planning the most appropriate and least invasive surgical plan. Due to differences in shunt flow rates, vascular anatomy, and cosmetic deformity, treatment strategies have been documented to be case dependent, including surgical excision, ligation, or endovascular embolization.^{3,4}

Several studies demonstrate different presentations, diagnostic methods, and management options. However, cerebral arterio-venous malformation is a relatively rare intracranial disorder.⁵ In this study, we report a rare case of cerebral AVM in an adolescent- adult patient who experienced headaches and seizures.

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CASE REPORT

A 24-year-old woman who was referred from the Demak City Regional Hospital undergoing treatment at the Sultan Agung General Hospital, Semarang City with complaints of weakness in the right limb, atypical progressive chronic headache with VAS 8-9. No aura found. Based on alloanamnesis from the patient's family, the patient previously had seizures at home. The seizure lasted approximately 5 minutes. When a seizure occurs, the patient's body contorts, the eyes bulge, and foam comes out of the mouth. After the patient experiences a seizure, the patient vomits and loses consciousness. After the seizure, the patient was taken to the emergency department of the nearest hospital. 5 years ago, patients often said that the throbbing headache seemed to come and go suddenly. Initially the pain appeared in the left occipital and cervical parts of the head and then centered in the frontal area. Headaches also appear on alternating sides, such as the right/left temporal part, as well as the lower part of the head. The patient can still carry out activities if an attack occurs. To relieve complaints of headaches, patients usually just lie down and fall asleep. The patient had no previous history of trauma, infection, hypertension, diabetes, or stroke.

At the time of admission to our hospital, the patient's general condition appeared weak with GCS E3-V.aphasia-M6 with motor aphasia. The NIHSS (National Institutes of Health Stroke Scale) score of the patient is 12. On physical examination, the patient's vital signs were normal, there was no anemic jaundice, cyanosis, dyspnea, or abnormal skin pigmentation. Further examination showed no bruits in the cervical area and no palpable lymph nodes. Examination of the thorax, abdomen and musculoskeletal areas was within normal limits. Neurological examination revealed motor and sensory deficits in the form of hemiparesis rightside, paresis N. Complete blood test results showed: Hb: 13.3 g/dL, leukocytes 11,300/uL, erythrocytes 4.2 million/uL, platelets 359,000/uL.

A higher function assessment examination is carried out. The examination is assisted using writing tools. Examination of the patient's level of consciousness is fully conscious with the Glasgow coma scale (GCS) E4 M6 V. Aphasias value. Good orientation check. Aphasia examination revealed that the patient was unable to speak spontaneously, unable to name the object being pointed to and unable to repeat verbally, but was able to understand the examiner's commands. In apraxia examination, the patient was able to blow a lighted firecracker. In visual and tactile examination of agnosia, the patient is able to recognize and name objects and people around him. Immediate memory examination, new and visual memories were within normal limits. (Figure 1)



Figure 1.a

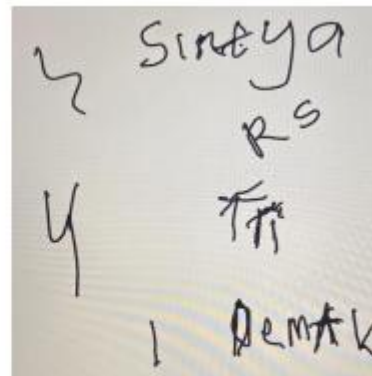


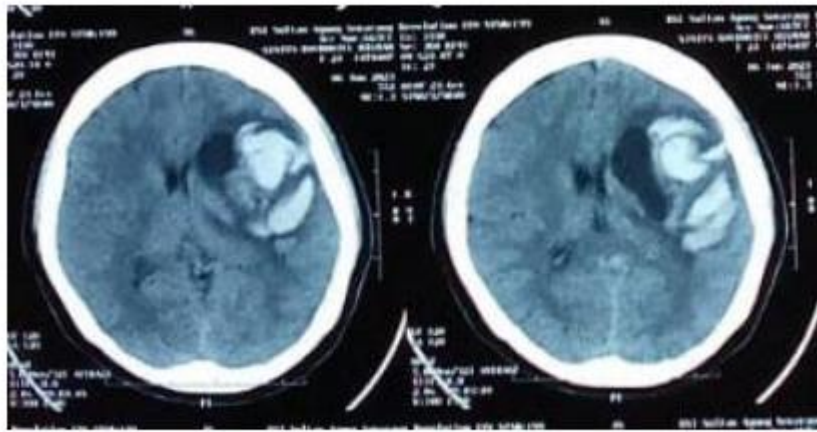
Figure 1.b

** Figure 1.a: patient closes eyes on command; b: patient writes answers to examiner's questions*

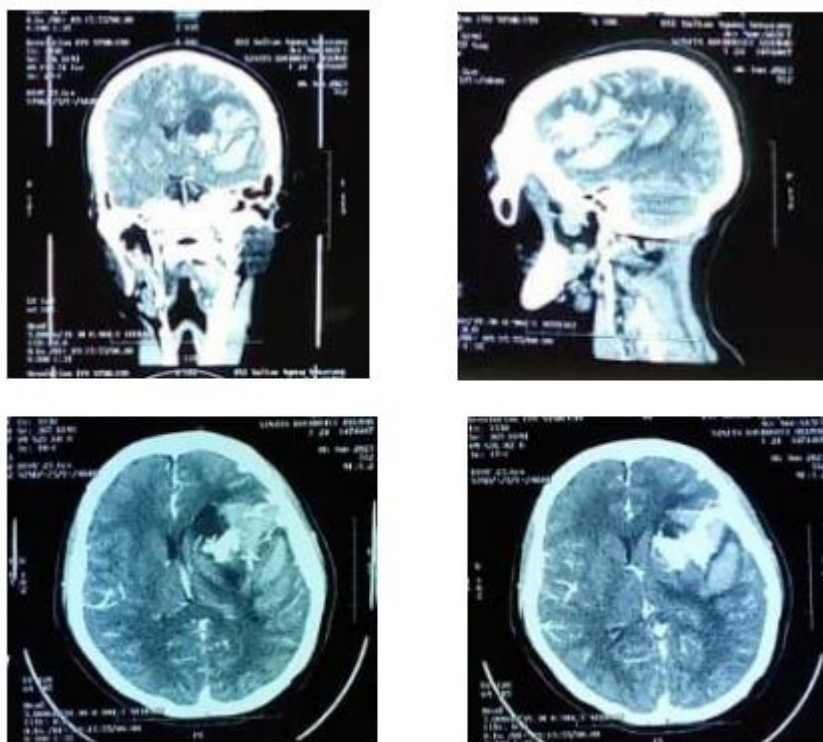
CT-Scan imaging of the head without contrast revealed intracerebral bleeding in the left temporal lobe with a volume of around 63.01 cm³ accompanied by perifocal edema, the left lateral ventricle appeared compressed, and the midline deviation to the right was >5mm. (Figure 2)

Meanwhile CT Angiography imaging showed a lesion resembling a bag of worms with a nidus measuring approximately 2.1 x 3.1 x 2.7 cm³ in the left temporal region, feeding arteries from the left middle cerebral artery and draining veins from the left transverse sinus. (Figure 3) An AVM was found in the left temporal region with a nidus size of 2.1 x 3.1 x 2.7 cm³ with a feeding artery from the left medial cerebral artery segment M-2 and a draining vein from the left transverse sinus with perifocal edema.

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**Figure 2. Non-contrast CT scan imaging*



**Figure 3. CT Scan angiography brain imaging*

The patient was suspected with left temporal BAVM. The patient was managed conservatively. Surgical resection by neurosurgery was not performed due to consideration of the bleeding location which is possible for recurrent bleeding. The patient was referred for further vascular neurointervention. Integrated care between health facilities has an important role in patient recovery.

DISCUSSION

Below we present a case report of a woman with spontaneous bleeding due to congenital BAVM (brain arteriovenous malformation) which was manifested by headache, hemiparesis, decreased function (motor aphasia) and followed by decreased consciousness. BAVM is very rare with an estimated prevalence of 0.01%-0.05% in the general population. BAVM occurs equally in men and women and generally appears in patients aged 20-40 years. Atypical headaches, such as in the following cases, have a clinical presentation of around 5% -14%.⁵

Arteriovenous malformation or AVM is a congenital abnormality that can occur in the brain or spinal cord, formed from an abnormal network of arteries and veins connected by one or more fistulas. The genetic transmission pattern of AVM is not yet known. AVM is a common genetic disorder, although at least in the specific context of hereditary syndrome.⁶ The most common

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problems associated with AVM are headaches and seizures, where at least 15% of the population do not show any symptoms. Other symptoms that are often found include vertigo, pulsing in the head with aura or without aura, progressive deafness and decreased vision, confusion, dementia and hallucinations.

AVMs can cause neurological disorders through 3 mechanisms. First, through bleeding that occurs in the subarachnoid space, intraventricular space and brain parenchyma. Second, if there is no bleeding, seizures can occur in 15-40% of cases. Third, progressive neurological deficits may occur in 6-12% of patients over several months to several years. Slowly progressive neurological deficits are thought to be related to blood flow being sucked away from adjacent brain tissue ("steal phenomenon").⁷ CT scanning is used to identify areas of acute bleeding, and the results can suggest vascular malformations, more clearly if contrast is used. In addition, CT scanning can depict vascular calcification associated with AVM. Intracranial AVM therapy modalities include; observation and medication; surgery, which is the gold standard therapy; endovascular embolization; radiosurgery; and a combination of all of that. However, surgical resection alone is sometimes not sufficient to treat higher grade AVMs and those located in eloquent structures.^{6,7} Some endovascular modalities such as embolization are useful in reducing the flow of larger AVMs to facilitate later surgery. This can relieve the patient's symptoms and reduce the size of the lesion to increase the effectiveness of further therapeutic approaches such as surgery or radiosurgery.⁸

Although embolization alone generally cannot treat AVMs in a complex manner, previous reports have suggested radiological healing in 10%-40% of cases. This chance of success is greater in smaller AVMs. However, the risk of complications remains, reported to occur in as high as 26% of cases. This is related to the embolization procedure itself (vessel perforation, hematoma) or related to the materials used (pulmonary artery embolism causing pulmonary edema, bronchospasm due to the use of dimethyl sulfoxide solvent).⁹

These findings have proven that children and young adults are at higher risk for BAVM rupture. This may be due to differences in brain anatomy and biochemistry resulting in more fragile or more aggressive AVMs. Larger nidus tend to be more symptomatic but less likely to bleed than smaller nidus, and have a higher risk of causing epilepsy or headaches.¹⁰

BAVM with hemorrhage has a moderate prognosis. The risk of developing a first hemorrhage is lifelong, increasing with age (2-4% per year, cumulative). Most will cause symptoms throughout the patient's life. Spontaneous recovery is very rare (< 1% of cases), 75% are small lesions (< 3cm) with a single vein and 75% have spontaneous bleeding.¹¹ Appropriate and comprehensive management has an effect on the patient's recovery.

AUTHOR'S STATEMENT

The author declares that there were no conflicts of interest during the process of preparing the manuscript.

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