




# Cerebral Arteriovenous Malformation in a Pediatric Patient with Severe Systemic Hypertension

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## Abstract

### Keywords

- cerebral arteriovenous malformation
- severe systemic hypertension
- left ventricular hypertrophy
- pediatric
- general anesthesia
- perioperative complications

Arteriovenous malformations (AVMs) are a tangle of blood vessels that connects arteries and veins in which the feeding arteries are directly connected to the venous channels. Cerebral AVM needs special consideration due to high risk of bleeding and neurological deficit associated with morbidity and mortality. In pediatric population, prevalence rate of cerebral AVM is 0.02%. The systemic hemodynamic changes are seen more frequently with peripheral AVM, whereas its association with cerebral AVM is rare. We report a rare case of cerebral AVM in a 12-year-old child with severe systemic hypertension and electrocardiogram changes of biventricular hypertrophy planned for surgical resection of AVM in view of intracerebral bleed. This case illustrates the importance of detailed cardiac, renal, and endocrine evaluation in children with cerebral AVM with systemic hypertension and left ventricular hypertrophy to rule out other causes of systemic hypertension prior to surgery.

## Introduction

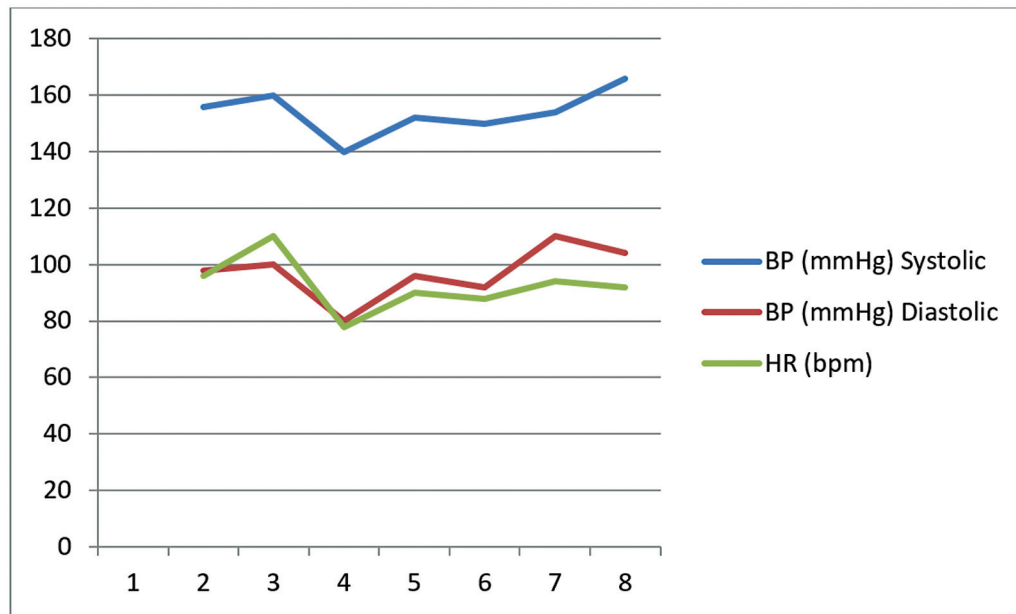
Arteriovenous malformations (AVMs) is a congenital vascular anomaly associated with direct connection between an artery and a vein without intervening capillaries which leads to hypertrophy of arterial and venous components along with increased blood flow. Cerebral AVM is uncommon with an overall prevalence rate of 0.5 to 1%.<sup>1</sup> In pediatric population the prevalence rate of cerebral AVM is 0.02% and the annual incidence rate of symptomatic AVM is 1.1 per 100,000/year.<sup>2</sup> Although it is not common in childhood, but rupture of AVM is more common in paediatric patients as compared to adults leading to spontaneous intracerebral hemorrhage. Intracerebral hemorrhage and its sequelae are the most common presentation in children.<sup>3</sup>

The initial investigation at the time of presentation is contrast-enhanced computerized tomography (CECT) brain followed by magnetic resonance imaging (MRI) after the clot has cleared; however, the definitive investigation is digital subtraction angiography (DSA) to visualize the vessels supplying the cerebral AVM. Surgery is often the first-line treatment, whereas embolization and stereotactic radiosurgery can be useful adjuncts in the treatment of AVM.<sup>4</sup> The peripheral AV fistulas such as femoral and brachial may lead to systemic changes causing cardiomegaly and increase in cardiac output leading to high output cardiac failure unlike intracranial AVM which may rarely cause systemic changes and cardiac abnormalities.<sup>5</sup> Here, we are reporting a case of cerebral AVM in a child with severe systemic hypertension.

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**Fig. 1** Vitals recorded during perioperative period. HR, heart rate; BP, blood pressure.

## Case Report

A 12-year-old male, weighing 20 kg, was admitted in the department of neurosurgery with complaints of headache for 3 months, fever, seizures, vertigo, and syncopal attack for 1 month, decreased vision in the left eye, and diplopia for 15 days. There was no significant past medical history. He was born at term via normal vaginal delivery. There was no history of head injury, developmental delay, prior hospitalization, dyspnea at rest/exertion, or poor performance in school. No family history of neurological or cardiovascular disorders was found. Neurological examination was unremarkable with no sensory, motor, and higher mental function abnormalities. CT head showed 3 cm left frontal lobe hematoma (Spitzel Martin grade 2, S1E1V0). Whereas CT angiography was suggestive of cerebral AVM involving the left anterior cerebral artery and superior sagittal sinus.

Patient was planned for surgical resection of AVM in view of intracerebral bleed. During preanesthetic evaluation, patient was conscious and oriented, afebrile, and heart rate (HR) was 86 beats per minute (bpm). Blood pressure (BP) in the left upper arm was 198/136 mm Hg. Due to such high BP, further measurement was done in all the other limbs and similar elevated readings were observed (→ **Fig. 1**). On auscultation of chest normal bilateral vesicular breathing was heard without any added sounds. During cardiovascular examination S1 and S2 were heard with no added murmur. Laboratory investigations were within normal limits. As the patient reported with episodes of seizures electroencephalogram (EEG) was done, which was unremarkable. Ophthalmology examination and chest X-ray were also normal. Electrocardiogram (ECG) showed tall QRS complexes along with notched T wave in lead V<sub>2</sub>-V<sub>5</sub> suggestive of biventricular hypertrophy. Two-dimensional (2D) echocardiography (ECHO) revealed concentric left

ventricular hypertrophy (LVH) with mild left atrial dilatation, trace tricuspid regurgitation, trace pulmonic regurgitation, and left ventricular ejection fraction of 65%. Renal ultrasound showed grade I medical renal disease with staghorn calculus in the right kidney. Renal Doppler study was normal. Thyroid profile and 24-hour urinary metanephrines were within normal limits. Upon consultation with cardiologist, tablet amlodipine 5 mg once a day was started to control hypertension.

For premedication, tablet alprazolam 0.125 mg was given at night and morning of surgery, along with tablet amlodipine 5 mg. In the operation theatre, vitals were recorded before induction, BP in the right arm with supine position was 160/100 mm Hg, HR was 110 bpm, and oxygen saturation (SPO<sub>2</sub>) was 99%. Preoxygenation with 100% oxygen was done. Injection midazolam 1 mg and injection fentanyl 2 µg/kg were given intravenously. Vital parameters after 1 minute were BP = 140/80 mm Hg, HR = 78 bpm, and SPO<sub>2</sub> = 100%. Then, the patient was induced with injection etomidate 0.3 mg/kg intravenously and injection vecuronium 1 mg/kg intravenously. After endotracheal intubation, the BP was 154/110 mm Hg which was further managed by using injection propofol (10 mg aliquots). A triple lumen central line in the right subclavian vein and arterial catheter in the left radial artery were secured. Maintenance of anesthesia was done using 50% oxygen, 50% air, and 1% sevoflurane (1 Minimum Alveolar Concentration [MAC] was achieved). During intraoperative period, hemodynamic stability was maintained, volume-controlled mode of ventilation was utilized, and analgesia was maintained using injection paracetamol 15 mg/kg. Intraoperative blood loss was around 200 mL which was managed by adequate intravenous fluids and blood transfusion. Surgery lasted for 3 hours. Patient was extubated after complete reversal of neuromuscular blockade. Postoperative CT scan showed complete removal of AVM with no cerebral

edema or residual bleed. In the postoperative period, systolic BP was in the range of 150 to 170 mm Hg and diastolic BP was in the range of 100 to 110 mm Hg for which the dose of antihypertensive was revised to tablet amlodipine 10 mg in the morning and 5 mg in the evening. Patient was discharged after 5 days. Upon follow-up, antihypertensive medication was stopped after 3 months.

## Discussion

AVMs are seen due to the presence of arteriovenous shunting through a nidus of coiled and tortuous vascular connections which connect feeding arteries to draining veins without any intervening capillaries in between. This results in hypertrophy of arterial and venous components of AVM. It can occur in the cerebral hemisphere, brainstem, and spinal cord. Although the etiology of AVM is still unclear, but most likely it is due to persistence of primitive AV connection or due to de novo synthesis of new connection later in life. Majority of the AVMs are congenital and it is hypothesized that it mostly occurs during early embryogenesis around the third week of life. Cerebral AVMs are congenital in origin which can present at any age. The reported mean age of diagnosis is 31 years.<sup>6</sup> Approximately 20% of all symptomatic AVMs present before 15 years of age. Most common presentation in children is spontaneous intracranial hemorrhage which could lead to headache, seizures, focal neurological deficit, decline in cognition, mass effect, and steal effect (diversion of blood to AVM causing cerebral ischemia). In case of suspected AVM, CECT brain is the initial investigation which shows serpiginous areas of enhancement; MRI brain shows lattice work of signal void spaces and signs of ischemia. However, DSA is the definitive technique to identify the vessel supplying the AVM. If episodes of seizures are present, EEG is also done. For the treatment of pediatric AVM, conservative management is no longer done as there is high risk of rupture which could lead to morbidity and mortality. Although surgical resection is the standard treatment, microsurgical technology has made this mode of treatment the quickest and complete method in achieving obliteration. Stereotactic radiosurgery is a new modality to maintain AVM obliteration in deep-seated lesions which are not accessible by microsurgery. Endovascular embolization is another technique gaining popularity.<sup>2</sup>

In children, hypertension is observed with the prevalence rate of 3.5%.<sup>7</sup> Although primary hypertension is more common in childhood compared to secondary hypertension which is mostly due to renal and renovascular diseases. According to the American Academy of Pediatrics guidelines, BP  $\geq$  140/90 mm Hg is defined as stage 2 hypertension which can result in end-organ damage. Therefore, it mandates medical management with the target BP of  $<$  130/80 mm Hg.<sup>8</sup> 2D ECHO is required in children with symptomatic hypertension even after nonpharmacological modifications, stage 2 hypertension, and hypertension associated with diabetes or chronic renal diseases.<sup>9</sup> In hypertensive state, the autoregulation shifts to the right and therefore

any degree of hypotension would cause cerebral ischemia due to reduction in the cerebral blood flow (CBF).<sup>10</sup> Abrupt increase in HR and BP often lead to complications such as myocardial infarction, pulmonary edema, and cerebrovascular hemorrhage.<sup>11</sup> Shang et al conducted a meta-analysis of observational studies on nephrolithiasis and risk of hypertension and found positive correlation between the two.<sup>12</sup> In our case, the child had 25 mm stag-horn calculus in the right kidney which was left untreated, whereas antihypertensive medication was stopped 3 months postsurgery. This indicates that hypertension in our case was not related to the renal disease.

Fujishima et al found cerebral AVM as a cause of cardiac hypertrophy in adults. In their study, 5 out of 11 patients with increased CBF had ECG changes such as bradycardia, LVH, and Wolff-Parkinson-White syndrome. Cardiac hypertrophy such as LVH in these patients was not due to hypertension, arteriosclerosis, severe anemia, or renal disease which is suggestive of its relation to increased CBF secondary to AVM. The association of LVH with AVM could possibly be due to relative hypoxia to myocardium or diastolic overloading of the left ventricle. Unlike large peripheral AVMs, cardiovascular effects associated with intracranial AVMs are uncommon as the caliber of vessels is not large and the location of intracranial AVMs is higher than the heart which favors a lower shunt flow in the upright posture. Cerebral AVM being enclosed by the skull retards the growth of AVM and collateral channels. Cardiovascular disorders associated with cerebral AVM are congestive heart failure and LVH due to increase in stroke volume.<sup>13</sup> Several studies have been conducted to show the association of high output cardiac failure with AVM. Saavedra et al conducted a case report on reversible pulmonary hypertension and high output cardiac failure triggered by pregnancy in a patient with congenital AVM and found that pregnancy and AVM are independent factors for high output cardiac failure.<sup>14</sup> Hence, any patient with symptoms of high output cardiac failure should be evaluated for the presence of AVM. The cardiac manifestations of cerebral AVMs are mostly seen in adults, and we found similar cardiac and ECG changes suggestive of biventricular hypertrophy (Katz-Wachtel phenomenon) associated with severe systemic hypertension in a pediatric patient with cerebral AVM.

In our case, the child presented with congenital cerebral AVM and severe systemic hypertension with ECG changes of biventricular hypertrophy. So, he was thoroughly investigated using 2D ECHO for mapping congenital cardiac anomalies, renal Doppler scan to evaluate for renal and renovascular diseases, and 24-hour urinary metanephrines to rule out pheochromocytoma.

This case illustrates the importance of detailed cardiac, renal, and endocrine evaluation in children with cerebral AVM with systemic hypertension and LVH to rule out other causes of systemic hypertension prior to surgery.

## Conflict of Interest

None declared.

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