CASE REPORT

Tangled Throughout Life: A Case of Paediatric Ruptured Cerebral Arteriovenous Malformation

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ABSTRACT

Arteriovenous malformation (AVM) in children is uncommon occurrence defined as presence of arteriovenous shunting through coiled and tortuous vascular connections. We discussed a case of a 3-year old girl presented with acute left facial asymmetry and right-sided limb weakness. Neurological examination revealed MRC scale of 0 out of 5 for power on her right side. Magnetic Resonance Angiography (MRA) revealed bilateral thalamic AVM. Surgical resection was not advisable in view of deep-seated location. Paediatric AVM most often become apparent following rupture with majority presents with headache. Ruptured paediatric AVM carries high burden of morbidity and mortality. Paediatric intracranial haemorrhage posed tremendous concern regarding its long-term outcome. Treatment would be more appropriate sooner rather than later especially for those presented with ruptured AVM. Surgical resection remains the gold standard treatment for all accessible paediatric AVMs with embolization and radiosurgery as adjunctive therapies. AVM in paediatric population is rare but carries grim prognosis.

Keywords: Arteriovenous malformations, Intracranial haemorrhages, Cerebral haemorrhage, Paediatrics, Child

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INTRODUCTION

Cerebral arteriovenous malformation (AVM) is an uncommon occurrence with prevalence of only less than 1% in paediatric population (1). It is defined as presence of arteriovenous shunting through coiled and tortuous vascular connections formed by a mutation early in embryogenesis (2). Due to it being congenital abnormalities, often we are unaware of its presence until the symptom starts to emerge. Clinical presentation of cerebral AVM often becomes apparent after it ruptures, resulting in intracranial haemorrhage.

Surgical treatment is still the gold standard for accessible cerebral AVM. Other modalities such as radiosurgery and embolization serve as adjunctive therapy and used in a multimodal approach or sole therapy. We describe a case of a young child with ruptured cerebral arteriovenous malformation (AVM) presenting with devastating motor deficits. Radiological evaluation revealed deep seated AVM in which aggressive treatment by surgical removal carries high risk of poor outcome. Embolization therapy was planned, unfortunately she defaulted her follow up.

CASE REPORT

A 3-year old girl was brought to the emergency department of a suburban hospital with acute onset of right sided hemiparesis after waking up from sleep approximately two hours prior to her presentation. She was otherwise a healthy child and achieved appropriate developmental milestones.

At presentation, she was alert and her vital signs were within normal range with blood pressure of 92/56 mmHg, pulse of 112 beats per minute, respiratory rate of 18 per minute and saturation of 100%. Neurological examination revealed Medical Research Council (MRC) scale for muscle power of 0 out of 5 with hypotonia and brisk reflexes in both right upper and lower limb. There were noticeable signs of upper motor neuron lesion on 7th cranial examination with absence of right sided forehead wrinkle and loss of nasolabial fold. Multiple hemangiomas were seen over her bilateral lower limb largest measuring at 2 x 2 cm.

Her initial non-contrasted computed tomography (CT) scan of the brain showed left intra-parenchymal haemorrhage at the basal ganglia measuring 2.2 x 2.0 x 2.6 cm with midline shift of 0.5 cm (Fig. 1). Subsequent magnetic resonance angiography (MRA) displayed thalamic arterio-venous malformation (AVM) of 4.1 x 1.7 x 3.0 cm in size over the left thalamus. Diagnosis of

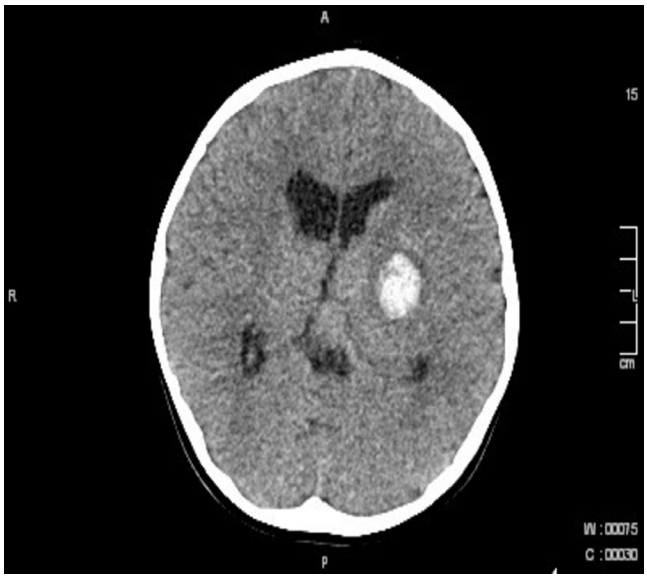

Figure 1: Non-contrasted CT brain showed intraparenchymal haemorrhage at left basal ganglia with perilesional oedema. There is effacement of frontal horn and body of ipsilateral lateral ventricle.

Spetzler Martin grade IV left thalamus AVM complicated with left basal ganglia bleed was made. An incident finding within the right thalamus was 2.3 x 1.2 x 1.7 cm size of another arterio-venous malformation (AVM). There were multiple small feeding arteries seen arising from supraclinoid portion of both internal carotid arteries and posterior communicating arteries (Fig. 2).

Referral to paediatric neurosurgeon from nearby tertiary hospital was made and was deemed inoperable in view of the deep-seated location with many feeder vessels. She was planned for possible embolization after digitized substraction arteriogram (DSA) in the tertiary hospital. Unfortunately, she defaulted her follow up ever since.

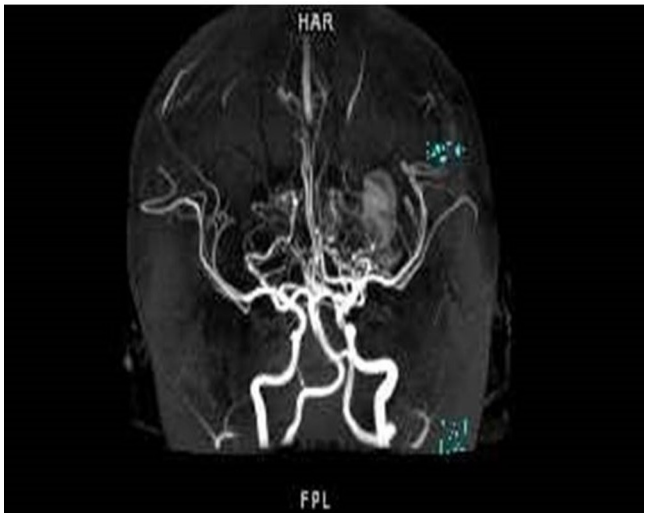

Figure 2: Magnetic resonance arteriogram/venogram showed multiple small feeding arteries arising from both internal carotid arteries with enlarged tortuous vein seen in both basal ganglia. Left basal ganglia haematoma is also seen.

DISCUSSION

Cerebral arteriovenous malformation (AVM) is an uncommon occurrence in the paediatric population. More recent population study of prevalence involving paediatric cerebral AVM is scarce. Prevalence of cerebral AVM in the whole population is less than 1%, more so in paediatric population (1). AVM is defined as presence of abnormal shunting through a nest of coiled and tortuous vascular connections bridging the arteries with the veins (2). Accurate pathophysiological event of the anomalous shunting is generally not well appreciated. It is understood that the pathology of AVMs started during embryogenesis where there is either failure of the embryonic AV connection to close or regrowth of a new vascular connection after closure of the primitive ones (2).

Although the pathology of cerebral AVMs is congenital, there are variations in the age of initial presentation. Age of diagnosis ranged between 1 to 18 years with mean age of 12 years (3). AVM in paediatric population often become apparent following rupture. Even though there are other causes of intracranial haemorrhage such as tumour, blood disorders and trauma, AVM is still the most common cause of brain haemorrhage in children with occurrence about 47.3% to 75% (1,3). Other clinical presentation of cerebral AVM includes seizure, neurological deficits and alteration in conscious level (4).

Since majority of children presented with intracranial haemorrhage at diagnosis, it is worthwhile to understand the consequences of this for children in the long term. Children with intracranial bleeding due to AVM has higher burden of morbidity and mortality (2). Several risk factors of haemorrhage in cerebral AVM has been reported. Deep location of nidus, size of nidus, a fast arteriovenous shunt, deep venous drainage and absence of generalized venous ectasia were observed to have statistically significant correlation with haemorrhage presentation for children with cerebral AVM (3). Association between AVM and haemorrhage are not only based on anatomic features of AVM. The inflow and outflow of the angioarchitecture correlate with risk of bleeding in paediatric cerebral haemorrhage population. Cerebral AVM with fast arteriovenous shunt and absent venous dilatation indicates a high inflow with higher resistance within the nidus and this type of AVM carries annual rupture rate as high as 11.1% (3).

Frequent initial imaging evaluation for cerebral AVM is computed tomography (CT). CT scan is useful in providing the location and size of hematoma within the brain. However, more thorough and comprehensive information on the vascular abnormalities are needed for diagnosis and treatment planning. Magnetic resonance imaging with angiography provide a non-invasive option to evaluate the vessels in detail. Digitalised Subtraction

Angiography (DSA) has been widely accepted as gold standard for investigating intracranial vascular lesions with superior spatial resolution and ability to evaluate dynamic blood flow through and around AVM even the small vessels (5). Nevertheless, the true architecture of the vessels may be interrupted by the hematoma if the study is done too early after cerebral haemorrhagic event (2).

The main aim of treatment is for complete eradication of arteriovenous malformations. The choice of treatment is best individualized to achieve maximum obliteration while minimizing neurological complications and can either be single modality or multimodality. Surgical resection of cerebral AVM still offers the highest percentage of obliteration despite the scarcity of evidence in paediatric population. However, not all cerebral AVM is suitable for surgical resection. Spetzler-Martin grading system is used to assess risk of neurological complication following a surgical resection. Factors observed to have unfavourable clinical outcome include coma at presentation, large cerebral haemorrhage >30m2, hydrocephalus, Spetzler-Martin grade >3 and deep venous drainage (4). Other adjunctive therapy such as radiosurgery and embolization serve to increase the rate of obliteration. These modalities can also be used solely in treatment of AVM where surgery considered too risky.

CONCLUSION

Cerebral arteriovenous malformation (AVM) in paediatric population is rare but carries high risk of mortality and morbidity. Majority of children with cerebral AVM presents with intracranial haemorrhage. If left untreated, there is risk of subsequent haemorrhage worsening neurological status and even lead to death. Treatment of choice for accessible cerebral AVM remains surgical resection while adjunctive therapies such as radiosurgery and embolization help increase obliteration rate.

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The authors declare that there is no conflict of interest exist.

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