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Management of Arteriovenous Malformations in Pediatric Population: about two Cases

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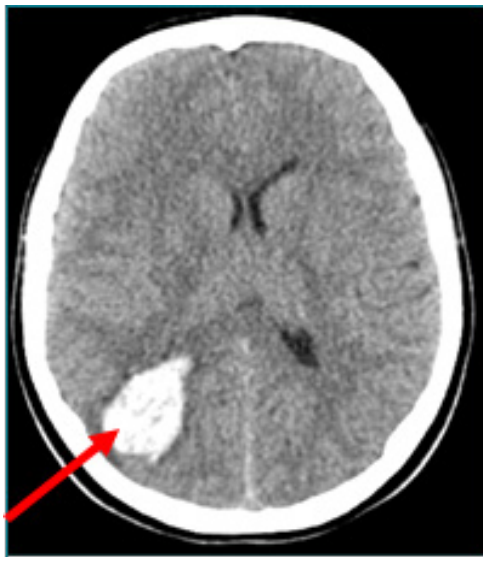
Introduction

The annual incidence of brain arteriovenous malformations (AVMs) in the general population is estimated between 0.1 and 4% with an annual hemorrhage rate between 2 and 10% and a 50% risk of neurological morbidity [1]. The re-rupture rate is estimated to be 2–4% resulting in a mortality rate up to 25% [1]. This risk is higher within the first 5 years after diagnosis. In comparison with the adult population, the literature regarding pediatric presentation is scarce. However, AVMs reportedly carry a higher rate of rupture in children than in the adult population. Several morphologic AVM characteristics are associated with hemorrhagic AVM presentation, including small AVM size, deep venous drainage, and the presence of associated arterial aneurysms. We report 2 cases of hemorrhagic presentation of brain AVMs in the pediatric population and their management.

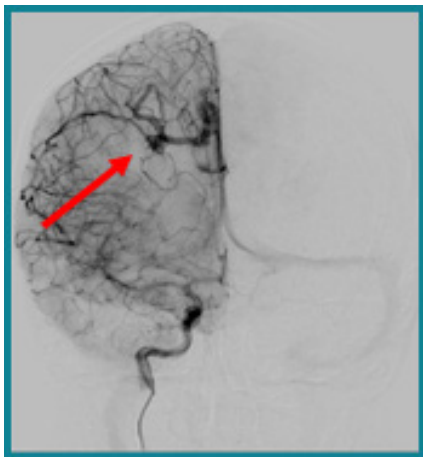
Clinical Case

Case 1: A 15-year-old girl was referred from another institution with acute unilateral headache, left hemianopsia, vision disorders and vomiting, without alteration of consciousness. Unenhanced brain CT showed a large right parieto-occipital hematoma. A cerebral digitalized subtraction angiography (DSA) was performed and confirmed the presence of an AVM. Embolization achieved the immediate angiographic cure of the lesion. At follow-up, the only residual symptoms are reading difficulties. Follow-up DSA at 3 months identified a tiny early venous drainage so that additional stereotactic radiosurgery by gamma knife was performed.

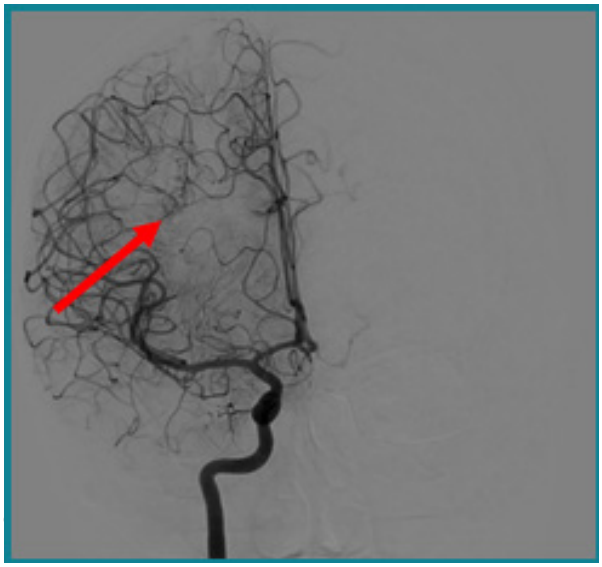
Case 2: A 11-year-old girl presented with acute onset of headache, vomiting and alteration of consciousness. Brain CT revealed a massive ventricular hemorrhage without edema. She was intubated and an external ventricular drain was placed. Brain MRI and DSA identified a deeply located brain AVM. Immediate embolization was performed, allowing to occlude 80% of the nidus and several AVM related aneurysms, suspected to be the cause of the hemorrhage. Additional gamma-knife radiosurgery has been planned as well. The patient keeps a discrete lower left limb paresis.



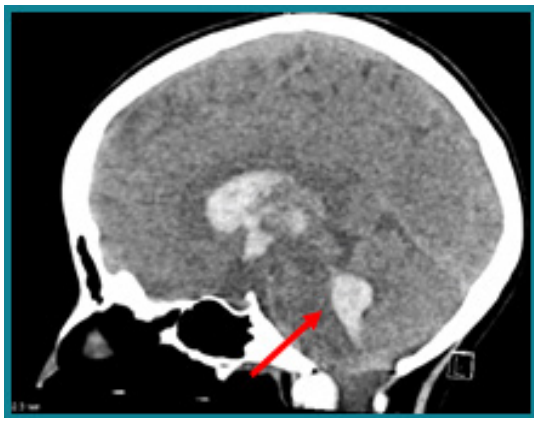
Enhanced brain CT at admission (axial MPR): right parieto-occipital hematoma



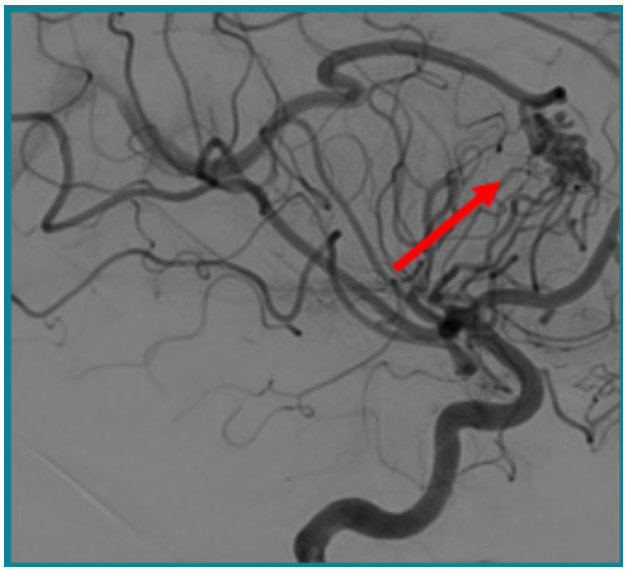
Pre-embolization DSA (frontal view) showing an early venous drainag



Post-embolization DSA (frontal view) showing complete angiographic obliteration



Unenhanced brain CT at admission (sagittal MPR): massive ventricular hemorrhage



Pre-embolisation DSA (oblique view) showing the nidus and the deep venous drainage

Discussion

AVMs are sporadic congenital abnormalities developing between the 3rd and 8th weeks of intrauterine life. They result from the persistence of a connection between one or several arteries and one or several veins without interposition of the capillary bed (Figure 1). In spite of their congenital origin, only 18- 20% of cerebral AVMs are diagnosed during infancy and childhood and are symptomatic after 15 years [1]. This could be due to the fact that most pediatric AVMs are only detected after rupture. Incidence represents 0.1 to 4% of the general population [1]. The prevalence is estimated to be between 0.06 and 0.11% [2].

Risk factors include the size (small), a previous history of hemorrhage, deep-seated or infratentorial AVMs, deep venous drainage, female sex, associated aneurysms, and diffuse AVM morphology. Only deep AVM location and exclusive deep venous drainage showed an independent effect on both initial and follow-up hemorrhage. Initial hemorrhagic presentation appears to be the strongest predictor for subsequent hemorrhage in untreated AVM patients.

The overall risk of hemorrhage from an untreated AVM in all age groups is estimated to be between 2 and 4% yearly, but the actual rupture risk may differ between distinct patient subgroups. They tend to rupture more frequently in children than in adults [1].

The symptoms at the time of presentation are varied. Intracranial haemorrhage is the presenting clinical manifestation in 75-80% of pediatrics patients [2]. Symptoms of congestive heart failure (18%) predominate in the newborn whilst neurological symptoms such as stroke, seizures or hydrocephalus (36%) occur more commonly in infants and older children [2]. The diagnosis is based on DSA.

Multimodality therapies are currently available. In most cases, the complete cure requires several interventions. The Spetzler-Martin grading system classifies AVMs based on location, size, and draining venous system, and it is used to assess the patient's risk of neurological deficit after open surgical resection. Surgery is less often performed and reserved to cases where hematoma removal is necessary.

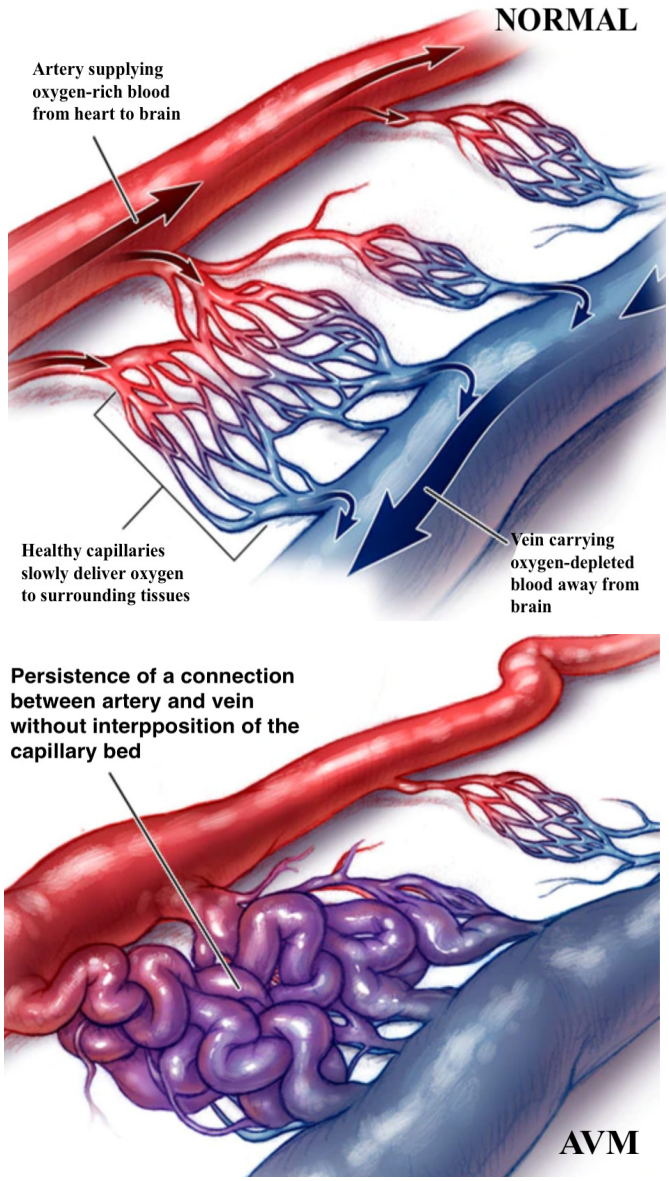


Figure 1: Embryogenesis of AVM

One or several trans arterial or rarely transvenous embolizations reduce the risk of bleeding and is associated with another therapy in case of AVM remnant, most often stereotactic radiosurgery (gamma-knife). This strategy is efficient and associated with low morbidity and mortality rates. Recommendations promote early treatment intervention in patients presenting with AVM hemorrhage, particularly in those harboring additional morphologic risk factors.

Conclusion

Upon initial diagnosis of intracerebral hemorrhage on noncontrast CT, workup and treatment should be initiated without delay. MRI and arteriography should proceed in the management. Intensive care and monitoring are indicated. Depending of the location of hemorrhage, if high intracranial pressure is present, an external ventricular drainage or an evacuation of the hematoma must be performed, possibly including removal of the AVM as well. In other cases, embolization reduces the risk of rebleeding. Stereotactic radiosurgery is possible mostly in case of AVM remnants after embolization. Prospective data on treatment-related AVM morbidity, however, are lacking.

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