Case Report

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Intraventricular Arteriovenous Malformation Mimicking Tumor: A Rare Presentation

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Abstract

Arteriovenous malformation is an abnormal connection between arteries and veins, bypassing the capillary system. Majority of AVM occurs in cerebral parenchyma and infrequently encountered in the lateral ventricle and foramen Monro. We report an intraventricular AVM in an 8 year old girl mimicking tumor.

Keywords: Intraventricular Tumor; Arteriovenous Malformation; Lateral Ventricle.

Introduction

Intraventriculararteriovenous malformations (AVM)are uncommon and represent 4% of AVM in child and 1% of AVM in adults [1,2]. The most common locations are lateral ventricle and foramen of Monro.We report an intraventricular AVM in an8 year old girl.

Case Report

An 8-year-old girl presented with history of headache and vomiting of 2 month duration. Neurological examination showed no neurological deficits. Non-contrast and contrast CT of the head revealed a well defined, hyperdense lesion with minimal contrast enhancement in atria and occipital horn of right lateral ventricle **(Figure 1 A & B).** MRI showed a well definedheterogeneously enhancing intraventricular lesion. Foci of bloomingwas seen on gradient echo image (GRE) **(Figure 1 C, D &E)**. A radiological impression of choroid plexus papillomaand the possibility of intraventricular meningioma werekept in mind and the patient was prepared for surgery.

A right parieto-occipital craniotomy (transcortical approach)withcomplete excision of the lesion was performed. Intraoperatively, a moderatelyvascular, grayish-blue, solid cystic, non friable lesion was identified in the right lateral ventricle. Small cystic component had hemosiderin deposit. Post operative recovery was uneventful and postoperative CT showed complete excision of the lesion **(Figure 2 A)**. Histopathological examination (HPE) of the specimen showed blood filled variable (medium to large) tissue, vascular channels surrounded by fibrocollagenous tissue with mild mixed inflammatory infiltrate along with thickened blood vessel wall. Focal area of fibrillary glial tissue was also seen **(Figure 2 B)**.

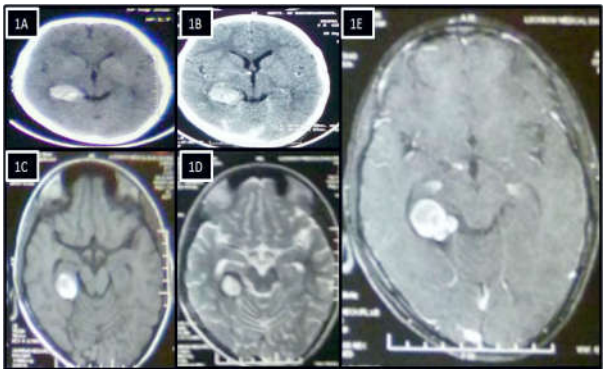

Figure 1: (A-B) Axial plain and contrast CT HEAD, **(C)** Axial MRI brain T1W1 images, **(D)** MRI brain T2W1, **(E)**MRI brain contrast showing lateral ventricle mass

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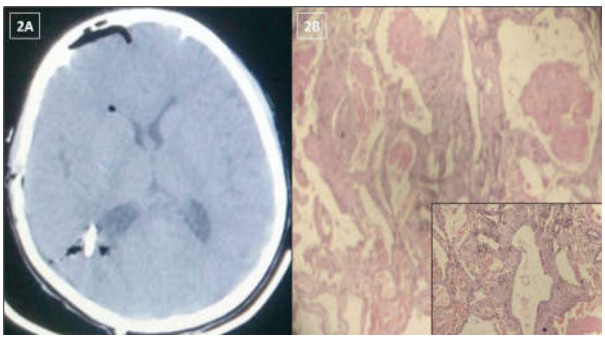

Fig. 2: (A) Post-op CT image with external ventricular drain, **(B)** 10X H&E stainshow dilated vascular channel. Few of them show arterialization of vein, (inset- 20X)- show dilated and congested vascular channel suggestive of arteriovenous malformation

Immunohistochemistry (IHC) was performed for confirmation; the tumour cells were strongly positive for CD34 and vimentin. Based on HPE and IHC, a diagnosis ofarteriovenous malformation (AVM) was established. Patient doing well at 6-month follow-up.

Discussion

Arteriovenous malformation is an abnormal connection between arteries and veins, bypassing the capillary system. Majority of AVM occurs in cerebral parenchyma and infrequently encountered in the lateral ventricle and foramen Monro[1].The clinical presentation of AVM in children is different from those in adults. There is a high propensity (80%) for the AVM childhood to present as haemorrhage in comparison to adults [1-5]. Likewise, epilepsy was reported in 12-18% of the AVM's children series and in 16 to 53% of the adult patients[6-9]. In neonates, AVM has been recognized as a cause of life-threatening congestive heart failure[10]. Mostly intraventricular AVM have been present with spontaneous hemorrhage into the ventricle, and this causes post-hemorrhagic hydrocephalus via blockage of CSF absorption or mechanical blockage of CSF pathways. Hydrocephalus due to aqueductal lesion has also been reported [11].

Radiological findings of AVM may be characteristic but are not pathognomonic.AVM have high signal attenuation on CT scans and showed intense enhancement. CT may be better than angiography in detecting angiographically occult intraventricular AVM [11]. CT scan also helps in recognition of the extent of the hemorrhage or the degree of hydrocephalus accompanying the AVM. On MRI, the nidus is hypointense on T1W1 images and hyperintense on T2W1 images. The signal void linear straits belonging to drainage vessels seen on MR angiography gives a clue to the existence of AVM. MRI can thus be helpful in differentiating highly vascular neoplastic lesions such as choroids plexus papilloma and carcinoma[12]. Cerebral DSA is the most reliable radiological investigation for the diagnosis of AVM. Intraventricular AVMs are commonly angiographically occult, because these lesions are often too small to be detected by angiography or because hemorrhage or thrombosis of the involved vessels may destroy them[13,14].Correct preoperative diagnosis of intraventricular AVM is important for appropriate treatment planning.Misdiagnosis is common because the characteristic signal void may be absent or angiographic results may be negative. Moreover, some intraventricular AVM may radiologically mimic tumors (as in our case) [11,15].

Despite the fact that intraventricular AVMs are often inoperable because of their deep location and intimidating vascular patterns, the natural history is unfavorable with a high incidence of hemorrhagic complications, and hence an intraventricular AVMs must be treated [16]. Surgical excision, endovascular embolization, radiosurgery or a multimodality approach have been used to treat this condition, however studies are not conclusive yet [17]. The ideal treatment for a cerebral AVM is total surgical resection. Total surgical excision is very important for intraventricular AVM because any mass left after surgery, radiosurgery or embolization can result in hemorrhage or hydrocephalus. AVM of the ventricles aregenerally small enough to be removed safely by microsurgical techniques alone[18]. Preoperative embolization may be helpful, although embolization is rarely curative. Yamada et al., reported the endoscopic resection of the intraventricular AVM [19]. Fahim et al., reported that the transtubular microendoscopic approach may beadvantageous for resecting intraventricular lesions by avoiding unnecessary retraction; therefore, it may reduce the risk of injury to the surrounding brain tissue [20]. Microsurgery is safer in managing hemorrhagic complications arising during surgery as compared to neuroendoscopy [21].

Several authors have reported poorer prognosis in children with AVM in comparison to adults[6,7].Conversely, a better prognosis was suggested for purely intraventricular haemorrhage AVM by some reports [22,23].

Conclusion

Although intraventricular AVM are very rare lesions, a higher propensity to bleed mandates aggressive management of such lesions. The ideal treatment for an AVM is total surgical resection. Generous reporting of usual lesion at unusual locations would helpin better understanding of lesions.

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