A rare case of dural arteriovenous fistula presenting as primary intraventricular hemorrhage

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ABSTRACT
Primary intraventricular haemorrhage (PIVH) is rare. Dural arteriovenous fistula causing PIVH is extremely rare. We report a case of a 17 year old boy who presented with left hemiparesis, left lower motor neuron facial palsy and ataxia. His computed tomography head revealed primary intraventricular hemorrhage. Catheter super selective angiography revealed a dural arterio venous fistula with arterial feeder arising from the middle meningeal artery as well as from the inferior marginal tentorial artery. Glue injection led to successful disappearance of the fistula and eventual clinical recovery.

Key words: Arterio-venous fistula, catheter angiography, cortico-venous reflux, primary intraventricular hemorrhage, venous sinuses

Introduction
Primary intraventricular hemorrhage (PIVH) was first defined by Sanders[1] and described as bleeding in the ventricular system without a discernable parenchymal fragment.[1-5] PIVH is considered as rare and constitutes 3% of all the spontaneous intracranial hemorrhages.[1,5,6] Hypertension is one of the most commonly associated risk factors for PIVH,[4,5] but it has been described with arteriovenous malformations (AVMs), aneurysms, moyamoya disease, coagulopathy and arteriovenous fistula.[3,5,7] PIVH is a rare presentation of a dural arteriovenous fistula (dAVF).[7-10] dAVF is vascular malformations that consist of an AV shunt in the wall of a dural sinus without an intervening nidus.[7,11,12] Purpose of presenting this case is to emphasize that all patients with PIVH should undergo catheter angiography to find the etiology, and condition like dAVF though rare is potentially treatable.

Case Report
A 17-year-old male presented with sudden onset severe headache, left sided weakness, swaying towards the left side, slurring of speech associated with right sided facial deviation and inability to close the left eye. There was no history of seizure, unconsciousness, vertigo, diplopia or dysphagia. On examination, patient was conscious and oriented. His vitals were stable. Neurological examination revealed left lower motor neuron type of facial palsy, pyramidal weakness of the left side in the form of spastic hemiparesis (power 4/5) with brisk reflexes and extensor plantar. Cerebellar examination revealed finger-nose incoordination on the left side and ataxia.

His blood hemogram and biochemistry tests including coagulation parameters were normal. Noncontrast computed tomography (NCCT) head revealed blood in all the ventricles, predominantly involving the fourth ventricle [Figure 1a] and occipital horn of the lateral ventricle [Figure 1b]. There was no associated intraparenchymal or subarachnoid bleed.

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Brain CT angiography appeared to be normal [Figure 1c] without any evidence of moyamoya disease, malformations or aneurysm. To identify the underlying etiology catheter angiography was planned.

Digital subtraction angiography images following left internal carotid artery (ICA), bilateral vertebral artery and left external carotid artery (ECA) injections were normal. However, RICA injection showed prominent inferior marginal tentorial artery arising from the cavernous segment of ICA giving filling to venous sinuses (transverse and sigmoid sinus junction) [Figure 2]. Right ECA injection arterial phase showed filling with the venous sinuses [Figure 3a] with cortical venous reflux [Figure 3b] and drainage into the transverse and sigmoid sinuses [Figure 3c] suggestive of cognards classification stage (II A + B). In order to find the main feeder of the fistula, a superselective angiography was done. Micro catheterization of the right middle meningeal artery (MMA) confirmed that the fistula was filling from the right MMA [Figure 4a]. The catheter was further advanced, and glue (N-butyl-2-cyanoacrylate [NBCA]) was injected with lipidol (50/50 ratio) [Figure 4b] following which the fistula, as well as the corticovenous reflux, disappeared [Figure 4c]. Glue injection led to successful disappearance of the fistula and eventual clinical recovery. Ataxia and facial weakness recovered over next 1-month.

Discussion

Primary intraventricular hemorrhage is defined as bleeding within the ventricles of the brain without associated
parenchymal or subarachnoid hemorrhage demonstrated on NCCT.[8] They are usually associated with conditions such as moyamoya disease, hypertension, vasculitis and arteriovenous malformations (AVMs).[3,5,7] Intracranial dAVF is uncommon lesions. Their true incidence is unknown, although selected series suggest that they occur only one-tenth as frequently as intraparenchymal AVMs.[13]

The fistula represents an abnormal connection between the dural or the pachymeningeal branches of the cerebral arteries and dural veins. In addition, dilatation of the cortical veins may occur, predisposing the patient to intracranial haemorrhage.[14]

The venous outflow restriction causes retrograde diversion of the venous outflow to the cortical veins, then to deep medullary veins with resultant subependymal venous congestion and rupture of the congested fragile subependymal venous network causing intraventricular hemorrhage.[7,9] Cognard system is more detailed and elaborates on the direction of flow, whether normal (anterograde) or retrograde and the presence or absence of cortical venous recruitment.[15]

Thrombotic occlusion with subsequent recanalization of the dural sinuses is considered as the most common cause of a dural fistula.[7,10] dAVF accounts for 10-15% of all intracranial vascular malformations and most commonly occurs along the transverse and sigmoid sinuses.[7,13] dAVF can present with a wide range of clinical manifestations like stroke due to ICH, TIAs, seizures, and cranial nerve palsies.[7,10] ICH has been reported in up to 35-42% of dAVF and may be in the form of intraparenchymal, subarachnoid, subdural and intraventricular hemorrhage with various combinations.[13] However, dAVF presenting as PIVH is very rare and there are only few case reports. Halbach et al.[8] reported the first case of PIVH due to dural AV fistula in the transverse sigmoid sinuses. Irie et al.[9] reported a female who presented with headache and pulsatile bruit with PIVH due to dAVF. Kawaguchi et al.[10] too reported a case of PIVH with dAVF who presented with headache and vomiting. Though our patient had a headache, his focal deficits were probably due to impingement of the blood filled fourth ventricle on thepons. Treatment is by obliterating the fistula either by glue injection, platinum coils or both by transvenous or transarterial approach. Padmanabhan et al. successfully embolized using detachable platinum coils, which were placed within the distal transverse and proximal sigmoid sinuses until the retrograde cortical venous filling was obliterated completely. Kawaguchi et al. initially used liquid material transarterially, but coils were used later on due to incomplete occlusion. However, we obliterated the fistula successfully by NBCA glue injection.

To conclude, dAVF still remains a rare cause of PIVH, and one should consider catheter angiography in all patients presenting with isolated ventricular bleed. Early recognition and prompt endovascular intervention can prevent catastrophic rebleed.

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Conflicts of interest
There are no conflicts of interest.

References