Case Report

Surgical management of a pial arteriovenous fistula with giant varix in an infant

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A seven-month-old boy with a history of recurrent pneumonia since birth presented to us following a single generalized convulsion. The infant was born following a normal term, uncomplicated pregnancy. The developmental milestones were normal - including social smile, gripping of objects and sitting erect with head holding. On examination the blood pressure was 100/55, heart rate 133.

The head circumference was 41.5 cm. The anterior fontanel was abnormally small and nearly fused with prominent bulging of the sagittal, bilateral coronal and the metopic sutures. A loud biphasic bruit was audible over the entire left hemi-cranium, which became more pronounced and monophasic over the anterior fontanelle. There were no abnormal pulsations, proptosis or chemosis visible externally and no fundoscopic abnormalities. Neurologically, the child was alert and interactive with normal visual fixation, normal facial and limb movements and crying. The cardiac examination revealed normal first and second heart sounds, no cardiac murmurs and lungs were clear.

A computed tomography (CT) scan done with contrast revealed a large (5.1 cm), well-circumscribed, isodense space-occupying lesion in the left sylvian fissure, homogeneously enhancing with contrast [Figure 1]. The lesion was causing significant mass effect.

Key words: Cerebellar hemorrhage remote to craniotomy, infant, pial arteriovenous fistula, varix

Introduction

We wish to report a case of a high-flow pial arteriovenous fistula (AVF) with a giant venous varix presenting in the neonatal period. These rare lesions are of clinical importance since the natural history is associated with severe cardiac and neurological sequelae.[1-3] This child was successfully managed surgically and his neurological development has been good. The radiological features and surgical management are described.

Case Report

A seven-month-old infant presented with a generalized seizure. The radiological evaluation revealed a large arteriovenous fistula in the left Sylvian region. The fistula connected the left middle cerebral artery and the vein of Trolard. A giant varix was present at the venous end. The child underwent craniotomy, complete disconnection of the fistula and excision of the varix. Except for recurrent seizures, which were eventually controlled on anticonvulsants, the child's neurological development has been good on long-term follow-up of three years. Pial arteriovenous fistulae are rare intracranial vascular malformations. Though significant operative risks exist, they can be successfully managed surgically with good long-term prognosis.

Figure 1: Contrast-enhanced CT showing a giant varix in the left sylvian fissure. The probable arterial feeder and venous drainage are seen
and slight midline shift. A four-vessel cerebral angiogram [Figure 2] revealed a large arteriovenous fistula supplied by the distal left middle cerebral artery (MCA) draining through a tortuous vein of Trolard into the superior sagittal sinus (SSS). There was a large variceal dilatation of the proximal venous end measuring nearly 7 cm in length and approximately 5.5 cm in diameter. There was also significant dilatation of the occipital sinuses.

An echocardiogram showed a structurally normal heart with no signs of hyperdynamic state or cardiac failure. The chest X-ray showed no signs of cardiac failure or pneumonia. Visual evoked potentials showed normal response bilaterally. The hemoglobin level was 7.7 g/dl. All other remaining hematological and biochemical parameters were normal.

**Management:** The infant underwent a left frontotemporoparietal craniotomy. At surgery, a large pulsatile varix (~ 7 x 5 cm) was seen in the sylvian fissure [Figure 3], fed directly by the left MCA. Just proximal to the fistulous connection several pial branches were seen arising from the MCA stump. The varix coursed inferiorly to the middle cranial fossa floor, then turned upward and was continuous with a largely dilated vein of Trolard, draining into the middle portion of the SSS. Upon exposure, the dura was densely adherent along the coronal suture and torrential venous bleeding occurred from the vein of Trolard precipitating cardiac arrest. The infant was successfully resuscitated and hemostasis achieved. The MCA feeder was clipped using a straight aneurysm miniclip followed by temporary clipping of the venous outflow from the fistula. The fistula and varix were then excised in toto and the venous end ligated. The surrounding brain parenchyma rapidly became hyperemic and edematous. Hemostasis was achieved and the dural defect was repaired using the variceal sac.

Postoperatively, the child was conscious with no asymmetry of movements following extubation on the third day. A CT scan on the fifth postoperative day showed a large filling defect in the area of the varix excision [Figure 4]. There was diffuse hemorrhagic change along the cerebellar folia. The general condition improved and the child was discharged after 14 days. He had poorly controlled generalized seizures during the next three months that were eventually controlled on valproate and phenobarbitone.

A postoperative angiogram done after one month [Figure 4] showed complete obliteration of the fistula and reconstitution of the distal MCA. The subsequent developmental course was satisfactory; the child attained standing position at about eight months of age, walking independently by 22 months. Language development was slightly delayed. However, the child’s developmental milestones by three years of age, assessed using the Denver developmental screening test, including language development were all within normal limits. He has now joined school at age three and a half years. The head circumference was only 47 cm (< fifth percentile).

**Discussion**

Pial AV fistulae are rare lesions of clinical importance. The varix coursed inferiorly to the middle cranial fossa floor, then turned upward and was continuous with a largely dilated vein of Trolard, draining into the middle portion of the SSS. Upon exposure, the dura was densely adherent along the coronal suture and torrential venous bleeding occurred from the vein of Trolard precipitating cardiac arrest. The infant was successfully resuscitated and hemostasis achieved. The MCA feeder was clipped using a straight aneurysm miniclip followed by temporary clipping of the venous outflow from the fistula. The fistula and varix were then excised in toto and the venous end ligated. The surrounding brain parenchyma rapidly became hyperemic and edematous. Hemostasis was achieved and the dural defect was repaired using the variceal sac.

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The majority of AVMs occurring in the neonatal and infant period are vein of Galen malformations. Nongalenic cerebral AVM (or pial AVM) have been reported to account for roughly 22% of AVMs found in neonates and 35% of AVMs in infants. The common presentations in neonates are systemic cardiac manifestations (54%), seizures (31%), hemorrhage (15%). In infants, hemorrhage (38%), hydrodynamic disorders (38%), cardiac manifestations (16%) and seizures (8%) are known to occur. High-flow AVFs are much more common in neonates than in infants who generally harbor the nidus-type AVM.[1]

These lesions have recently been termed pial single-channel arteriovenous fistulae since they are composed of a single venous channel in communication with one or more arterial connections with no intervening nidus of vessels.[2] They account for only 1.6% of all AVMs and are associated with a poor natural history. Infants with these are known to develop hydrodynamic disorders that rapidly damage the surrounding brain. Subependymal or cortical atrophy, white matter calcification and delayed myelization have all been described in these children. Therefore, early intervention is essential for optimal neurological and cognitive development.[2,3]

In addition to standard microsurgical technique, additional measures such as induced hypotenion, temporary clipping and pharmacological neuroprotection may be helpful. However, preexisting venous hypertension may precipitate severe bleeding that does not respond to these and other standard hemostatic measures. Thus the aesthetic team should be ready in anticipation of severe blood loss and potential circulatory collapse. In this case, the varix was very superficial and could be easily excised following fistula disconnection. However, simple disconnection of the fistula should be the goal of therapy.[2] Attempts at excision of the varix may exacerbate parenchymal bleeding.

Hoh et al. have recently summarized the literature from 1970 to 2000. There have been 79 patients reported to date. Venous varices were found in 48 of 54 cases (89%). However, at that individual centre, only three out of nine were associated with varices. In their series, the concept of ‘flow disconnection’ either endovascularly or surgically was advocated.[2] Surgical disconnection involves either aneurysm clip application or cauterization of the feeding vessel. Though this has proven effective, some lesions are deep or surgically inaccessible and the risk of surgery can be very significant. As in this case, severe bleeding can be expected due to (1) venous hypertension due to the high-flow system and (2) hyperaemia from normal perfusion pressure breakthrough. Unlike the case of nidus-type AVM, the strategy of surgical disconnection without lesion resection was found to be sufficient for obliteration of the fistula. Hence, excision of the varix is probably not required.[2]

Obliteration of the fistula by an endovascular route, avoiding the risks associated with craniotomy, should always be considered especially when the lesion is deep seated or the risk of neurological deficit with surgery is high.[2,3,5] However, endovascular attempts are not always successful or safe. Several technical difficulties in obliteration of the fistula have been described. Endovascular embolization of the fistula may be complicated by migration of the embolization agent into a varix, to the lung or elsewhere in the cerebral vasculature.[2]

An additional interesting finding in this case was the occurrence of postoperative cerebral hemorrhage. Intra cerebral hemorrhage distant to the operative site is a rare complication of neurosurgical procedures. Both supratentorial hemorrhage following infratentorial surgery and cerebellar hemorrhage following supratentorial surgery are described.[9,10] In a recent review the incidence of distant cerebral hemorrhage following supratentorial craniotomy was approximately 0.3-0.6% with 98 cases being reported in the English literature.[10] Infratentorial hemorrhage remote to supratentorial procedures was most commonly associated with operations in the deep Sylvian fissure and para midial regions (81%).[9] Although not fully understood, one proposed mechanism of this phenomenon is that a transtentorial pressure gradient is set up by excessive CSF loss leading to disruption of the cerebellar venous blood flow and eventual venous hemorrhage. Fortunately, the sequelae are usually none or minimal, however, extreme vigilance is advised since neurological deterioration (14%) and even fatality (32%) have been reported.[9,10]

Pial arteriovenous fistulae with giant varix are rare vascular malformations that can be successfully managed surgically with good long-term prognosis. When surgery is undertaken, one should be alert to the risk of heavy venous bleeding associated with the venous hypertension of this high-flow system and the risk of postoperative hemorrhage due to normal perfusion pressure breakthrough phenomenon. The ideal management would be a concerted effort involving both the operating neurosurgeon and the endovascular interventionist.

References