A case of partial anomalous systemic venous drainage and perioperative detection of cerebral arteriovenous malformations

Nilanjan Dutta1, Debasis Das1, Raghu Maruti Govindappa1, Patralekha Das1, Rangan Koley2, Manish Kumar Sharma2, Amitabha Chattopadhyay3, Sanjiban Ghosh3
1Department of Cardiac Surgery, Narayana Superspeciality Hospital, Howrah, West Bengal, India, 2Department of Cardiac Anaesthesia, Narayana Superspeciality Hospital, Howrah, West Bengal, India, 3Department of Pediatric Cardiology, Narayana Superspeciality Hospital, Howrah, West Bengal, India

ABSTRACT

We report a case of 8-year-old boy with unexplained desaturation and clubbing. Echocardiography showed anomalous drainage of right superior vena cava into left atrium. He did not have any neurological symptoms preoperatively. Some perioperative observations and subtle postoperative behavioral changes prompted us to investigate him further. He was found to have extensive cerebral arteriovenous malformations and hemiatrophy of brain.

Keywords: Arteriovenous malformation, congenital, congenital heart disease

An 8-year-old boy was admitted to our unit with a history of mild exertional dyspnea. Clinical examination revealed the presence of cyanosis (room air saturation around 88%), clubbing and ejection systolic murmur with loud second heart sound.

A detailed echocardiography revealed right superior vena cava (RSVC) draining into roof of left atrium (LA). Interatrial septum was intact and there was mild atrioventricular valve regurgitation on either side. There was no other structural abnormality in heart. A contrast (agitated saline injected in the left upper limb) echo clearly showed immediate filling of LA with no opacification of the right atrium [Figure 1].

The patient underwent cardiac catheterization and computed tomography (CT) to rule out any other anatomical lesions. Catheterization study revealed partial anomalous systemic venous drainage of RSVC to the roof of LA. In addition, RSVC was markedly dilated [Figure 2] with one right upper pulmonary vein draining high up into it. Right-sided chambers were smallish and LA and left ventricle were dilated. CT pulmonary angiography also confirmed the same findings clearly mentioning significantly dilated internal jugular veins and neck vessels.

The patient underwent surgery by median sternotomy on cardiopulmonary bypass. RSVC was connected to right atrial appendage with an intervening pericardial tube constructed over 11 size Hegar dilator [Figure 3]. Azygos vein was divided to ease the mobilization of RSVC. LA end of RSVC was closed with a small piece of autologous pericardium. The right upper pulmonary vein was left draining into RSVC. However, the significant intraoperative observation was bright red blood coming through RSVC cannula, very high mixed venous oxygen saturation, and requirement of vasopressors to maintain adequate perfusion pressure. The patient was weaned off cardiopulmonary bypass with very high dose of norepinephrine (0.2 mics/kg/min).

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Dutta N, Das D, Govindappa RM, Das P, Koley R, Sharma MK, et al. A case of partial anomalous systemic venous drainage and perioperative detection of cerebral arteriovenous malformations. Ann Pediatr Card 2021;14:244-6.
The patient continued to be on high dose of vasopressors in intensive care unit, but his peripheral pulses were always bounding. Neck vessels were pulsatile too. He was extubated on day 2 after operation. The patient remained very irritable after extubation. His sensorium also deteriorated without any focal neurological signs or seizure. CT head and angiography brain were done. Plain CT head showed diffuse hemiatrophy of the right cerebral lobe with disorganized cortex. CT angiography of brain revealed hemiatrophy of right half of brain with large arteriovenous malformations (AVM) involving right cerebral hemisphere with arterial supply from right middle cerebral artery and draining into superior sagittal sinus [Figure 4a and b]. All cerebral arteries were dilated except bilateral anterior cerebral arteries. There was distinct opacification of venous sinuses in the arterial phase. All these findings were suggestive of cerebral AVM. Both neurological and neurosurgical opinions were taken. However, considering hemiatrophy of brain and widespread distribution of AVM, no interventional embolization was considered. The patient’s sensorium also improved significantly over course of stay in hospital. The patient was discharged with prophylactic antiepileptic (levetiracetam) on postoperative day 13. The patient’s family was prognosticated in detail from the neurological point of view, and advised neurology follow-up also. The volume overload from the one isolated pulmonary vein which was left draining to RSVC was managed with anti-failure medications and diuretics. The patient is in regular follow-up for 3 years with us and is doing well so far and is planned for a CT angiography in the next visit.

DISCUSSION

Congenital heart diseases are known to co-exist with other structural anomalies and brain is not an exception. AVM is most commonly seen in brain and spinal cord. The most common cause of cerebral AVM is vein of Galen malformation.\footnote{1} There is no specific cause of this...
malformation except that they tend to run in families and are more commonly seen in males. Most of the extensive cerebral AVMs present in neonatal or infantile period with intractable heart failure. Late presenters manifest with mass effects of AVMs such as ischemia, hemorrhage, and brain atrophy.

The patient presented to us at 8 years with cyanosis and clubbing due the partial anomalous systemic venous drainage. Although there is atrophy of the right cerebral cortex, there was no neurological symptoms preoperatively. The only thing which should have raised the suspicion was significant dilatation of R SVC and neck vessels in catheter and CT angiography. Another interesting finding was dilatation of left-sided chambers. Isolated drainage of R SVC to LA is a right to left shunt which should cause only desaturation, not volume overload. It is because of AVMs that the left heart chambers became dilated.

The purpose of presenting this case is to remind us of the possibility of cerebral AVM in presence of unexplained dilatation of upper body systemic veins, difficulty in maintaining adequate perfusion pressure during cardiopulmonary bypass and need for excessive vasopressor agents. These patients are also vulnerable to neurological complications perioperatively.

Author’s note
Consent for publication was granted by patient’s parents.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. McElhinney DB, Halbach VV, Silverman NH, Dowd CF, Hanley FL. Congenital cardiac anomalies with vein of Galen malformations in infants. Arch Dis Child 1998;78:548-51.
2. Hewitt AL, Morrical BD, Cetta F. Cerebral arteriovenous malformation detected by newborn congenital heart disease screen with echocardiography. CASE (Phila) 2017;1:242-4.