Systemic venous anomalies in a child with a vein of Galen

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ABSTRACT

We describe our findings in a child with a vein of Galen malformation, in whom the right superior caval and the azygos veins drained into the roof of the morphologically left atrium. A persistent left superior caval vein drained into the morphologically right atrium through the coronary sinus. The additional presence of dual brachiocephalic veins permitted the deployment of a multifunctional ventricular septal defect occluder device to occlude the right superior caval vein, correcting the right-to-left shunt. This also prevented azygos venous drainage into the left atrium.

Keywords: Brachiocephalic veins/abnormalities, caval vein, superior/abnormalities, embolization, therapeutic, vein of Galen malformations

INTRODUCTION

It is well established that although rare, drainage of the right superior caval vein to the morphologically left atrium can cause left ventricular overload and arterial desaturation. Surgical reimplantation is usually required to address this malformation. We recently encountered such a finding in a child with a vein of Galen malformation. The presence of a persistent left superior caval vein, draining through the coronary sinus to the morphologically right atrium, and with the additional presence of dual bridging veins, permitted us to occlude the right superior caval vein and avoid surgical reimplantation.

CASE REPORT

A 6-month-old boy (weight: 8.6 kg) underwent four sessions of endovascular treatment to occlude an aneurysm of the vein of Galen and was scheduled for further neurological interventions [Figure 1a-c]. The Galen vein was diagnosed by fetal ultrasound during antenatal congenital anomalies screening. After birth, the child underwent echocardiography as part of the work-up for anesthesia before the planned intervention on the vein of Galen. Transthoracic echocardiography examination revealed the heart to exhibit a usual atrial arrangement, with concordant atrioventricular and ventriculoarterial connections. There was persistent patency of the oval foramen. The right superior caval vein, along with the azygos vein, was found to be draining exclusively to the morphologically left atrium. There was a persistent left superior caval vein, which drained into the morphologically right atrium through the coronary sinus. The inferior caval vein also drained in a normal fashion to the morphologically right atrium. The left-sided chambers were significantly dilated at greater than the 2nd centile [Figure 2a-d and Video 1]. The child's oxygen saturation level was normal at 97%

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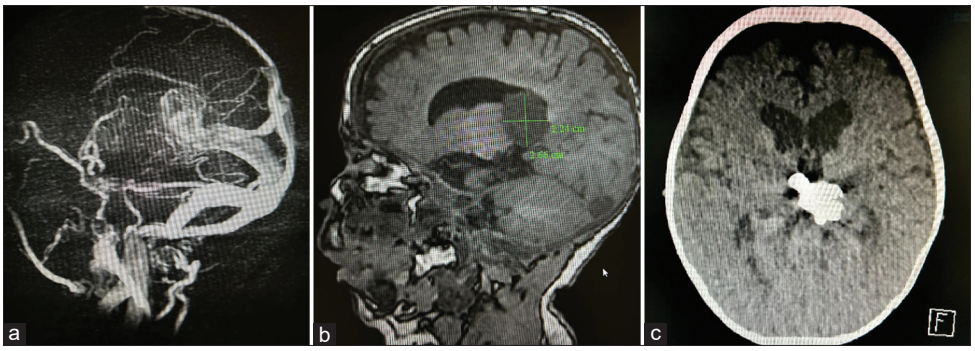

Figure 1: (a-c) The panels show the aneurysmal vein of Galen before (a and b) and after (b) by its embolization using coil-assisted glue copolymer

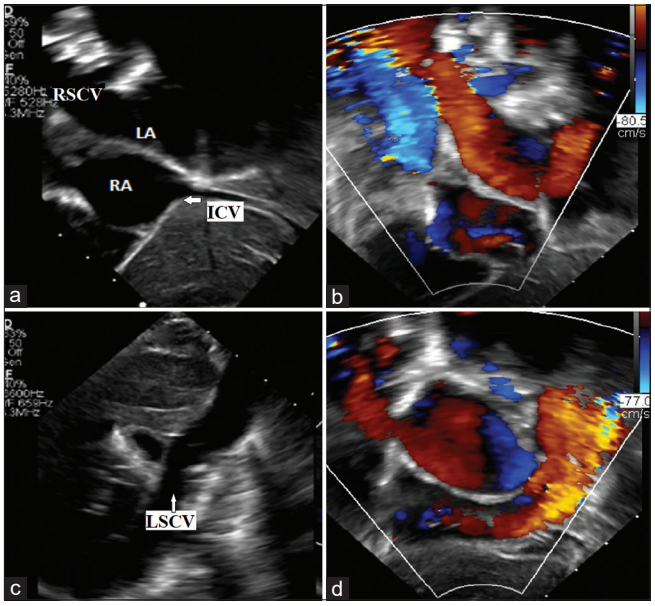

Figure 2: (a-d) Modified bi-caval (a) and modified high parasternal (b) transthoracic echocardiographic images with color flow mapping showing the right superior caval vein daring into the morphologically left atrium, with the inferior caval vein draining to the right atrium, (c and d) the left superior caval vein draining to the right atrium through the coronary sinus. RSCV: Right superior caval vein, LA: Left atrium, RA: Right atrium, ICV: Inferior caval vein, LSCV: Left superior caval vein

when breathing room air. However, on analysis of the arterial blood gas, it was found that the arterial partial oxygen pressure was only 43 mmHg. Due to a residual shunt through the vein of Galen and a left-to-right shunt due to the right superior and middle pulmonary venous return to the right superior cava vein, the child was in a high cardiac output state. This resulted in an oxygen saturation level above 90% despite the low arterial oxygen partial pressure. Three-dimensional computed tomography endocast and virtual dissection reconstructions demonstrated the right superior caval vein draining to the left atrium. There were anomalous connections of the right upper and middle pulmonary veins to just above the junction between the right superior caval vein and left atrium. The right lower and left pulmonary veins were connected normally to the left atrium with a bridging vein between the left and right superior caval veins [Figure 3a and b].

At age 4, cardiac catheterization was performed, and the angiographic images confirmed the previous findings. Contrast injection into the right internal jugular vein, however, revealed the presence of two bridging brachiocephalic veins, one closer to the aortic arch and another at the root of the neck lying anterior to the trachea, as well as the drainage pattern of the right and left superior caval veins [Figure 4a-c and Video 2].The dimensions of the right internal jugular vein, the right superior caval vein, the lower brachiocephalic vein, the left superior caval vein and the azygous vein were determined [Figure 4 d-f]. The mean right superior caval venous pressure was 11 mmHg. Measurement of the calibers of the venous pathways suggested that if the right superior caval vein entry to the left atrium were occluded, the two bridging veins would permit all the superior venous return to reach the right atrium through the left caval vein. Hence, we positioned a KONAR-MF ventricular septal defect occluder device (Lifetech, Shenzhen, China) partially in the azygous vein but mostly in the right superior caval vein at its junction with the left atrium, avoiding the orifices of the right pulmonary veins [Figure 5a and b]. An angiogram confirmed appropriate placement with no flow from the right caval and azygos veins to the left atrium [Figure 5c]. An echocardiographic assessment the following day confirmed the absence of any pericardial effusion. Both the chest radiograph and the electrocardiogram were normal. The child was advised to take aspirin for 6 months and was scheduled for routine follow-up. Transthoracic echocardiography was done 11 months after deployment of the device. The subcostal view showed a dilated coronary sinus, unobstructed flow of the right upper pulmonary veins, flow of left superior caval venous blood into the coronary sinus, good biventricular contractility, and a nondilated left ventricle. The apical four-chamber view showed a left ventricle of normal size with good contractility and a dilated coronary sinus. In the parasternal long-axis view, the coronary

sinus was found to be enlarged. The suprasternal view showed an open aortic arch, a well-positioned device, an unobstructed flow of blood from the right upper pulmonary veins, and a nondilated brachiocephalic vein with no turbulence [Video 3].

DISCUSSION

It is very unusual for the right superior caval vein to drain exclusively to the morphologically left atrium, particularly in the setting of an intact atrial septum. This lesion is even more unusual as a persistent left superior caval vein was draining through the coronary sinus to the morphologically right atrium. We encountered these unusual systemic venous malformations in a child who was undergoing neurological interventions for treating an aneurysmal vein of Galen.

The unusual combination of a right superior caval vein draining into the left atrium in an infant with a vein of Galen malformation was reported by Relan *et al.*[1] These authors performed agitated saline contrast echocardiography to confirm the diagnosis of the drainage of the right superior caval vein into the left atrium.[1]

The significant finding during the transthoracic echocardiographic interrogation of our patient was the dilated left heart. A detailed examination by color flow mapping revealed the right superior caval vein draining into the left atrium. Contrast echocardiography would have been helpful in this situation, but it was not performed on our patient. Instead, the child underwent computerized tomographic angiography, as there was a suspicion of an anomalous systemic pulmonary venous connection. The unexpected presence of yet another systemic venous anomaly, namely two large bridging brachiocephalic veins, permitted us to treat the child by occluding the right superior caval vein at its junction with the morphologically left atrium, avoiding surgical reimplantation.

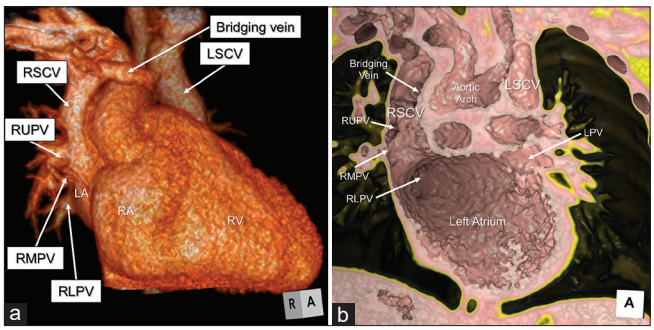

Figure 3: (a and b) Three-dimensional computed tomography endocast (a) and virtual dissection (b) reconstructions show the right superior caval vein (RSCV) draining to the left atrium (LA), anomalous connections of the right upper and middle pulmonary veins just above the junction between the RSCV and LA. The right lower and left pulmonary veins were connected normally to the LA. There was a bridging vein between the left superior caval vein and the RSCV. RA: Right atrium, RV: Right ventricle, RSCV: Right superior caval vein, LA: Left atrium, LSCV: Left superior caval vein, RUPV: Right upper pulmonary veins, RMPV: Right middle pulmonary veins, RLPV: Right lower pulmonary veins, LPV: Left pulmonary veins

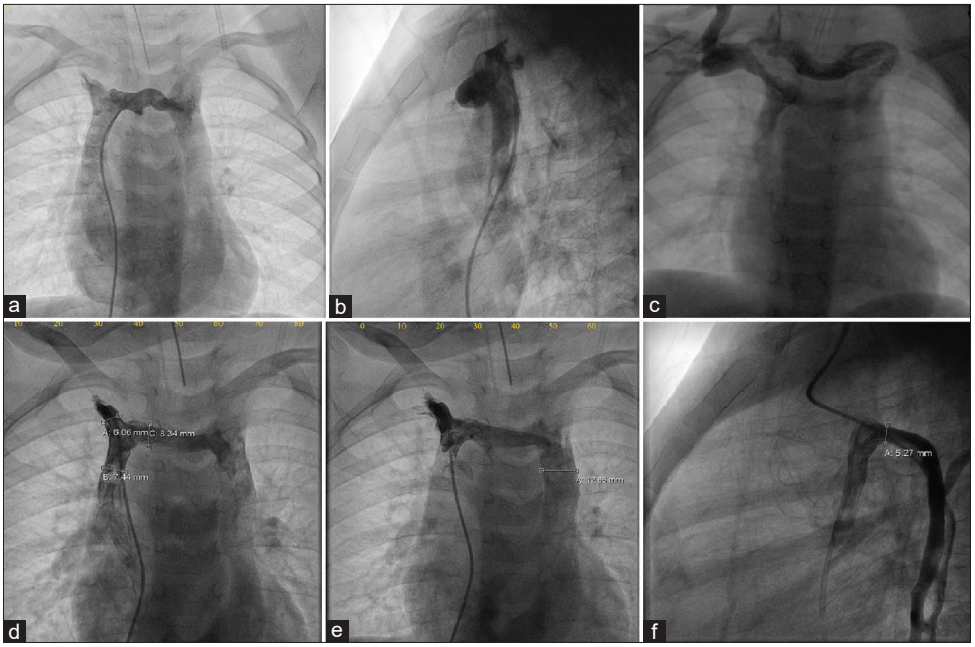

Figure 4: (a-f) Angiography shows the drainage of the right superior caval vein into the posteriorly placed left atrium (a), with the left superior caval vein draining via the coronary sinus into the right atrium (b). Paired brachiocephalic veins arose from the right internal jugular vein, one closer to the aortic arch and the other at the root of the neck (c). (d through f) the dimensions of the right internal jugular vein at 6.06 mm, the right superior caval vein at 7.44 mm, the lower brachiocephalic vein at 8.34 mm, the left superior caval vein with a width of 12 mm and length of 18 mm, and the azygous vein at 5.3 mm

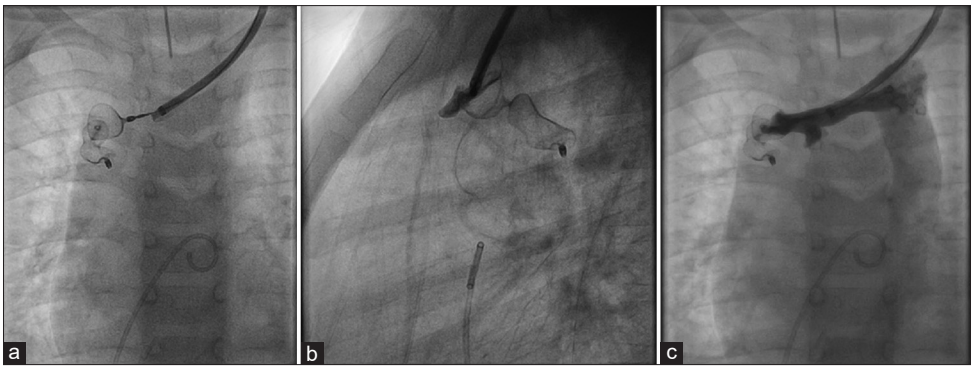

Figure 5: (a and b) Follow-up fluoroscopy showing the location of the occluding device relative to the azygous vein, mostly positioned in the orifice of the right superior caval vein (a and b). A venogram taken subsequent to deployment shows no flow into the left atrium (c)

It is well established that, by its complex embryological development, the brachiocephalic vein can show various arrangements, including an atypical superior location and duplication.[2-6] In our patient, the brachiocephalic veins joined anterior to the trachea, and no structures were trapped between them. We believe that it is unlikely for the brachiocephalic veins to cause airway compression, as they are soft vascular structures and likely to elongate as the child grows. The latest transthoracic echocardiography showed that the brachiocephalic veins were not dilated.

The success of our procedure depended on the accurate placement of the device chosen to occlude the right superior caval vein. If placed in the proximal part of the vein, above the entrance of the azygos vein, the azygos vein would be left draining to the left atrium. This would have permitted ongoing hypoxemia and overload of the left ventricle. If the device had been placed below the entry of the azygos vein, however, the funnel shape of the caval vein may well have promoted its embolization. A further potential problem was to place the device so as to avoid the openings of the right pulmonary veins to the left atrium. This was the most important aspect in our patient since, almost certainly, despite the integrity of the atrial septum, the caval vein could drain to the left atrium because of the initial presence of a superior sinus venosus defect.[6] This likelihood was confirmed by the virtual dissection of the computed tomographic dataset showing the right pulmonary veins draining into the right superior caval vein at its entrance to the left atrium. The multifunctional occluder device, therefore, was placed with one disc in the azygos vein, and the other in the right superior caval vein. We assumed that the azygos vein drainage would be through its communication with the hemiazygos vein. The disc in the azygos vein supported the device, preventing its detachment, while the other blocked the orifice of the caval vein. The unusual combination of the systemic venous anomalies permitted our successful cardiac intervention. Informed written consent from the parents of the child and the institutional ethical committee approval (CR#2023/23) was obtained to publish this case report.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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