2022

Right Superior Vena Cava Connected Left Atrium with Partial Anomalous Pulmonary Venous Return and Intact Atrial Septum: An Unusual Cause of Paradoxical Embolism

Follow this and additional works at: https://www.j-saudi-heart.com/jsha

Part of the Cardiology Commons

This work is licensed under a Creative Commons Attribution-Noncommercial-No Derivative Works 4.0 License.

Recommended Citation
Al-Sehly, Abdullah; Hrfi, Abdah; and BinSalahuddin, Ahmed (2022) "Right Superior Vena Cava Connected Left Atrium with Partial Anomalous Pulmonary Venous Return and Intact Atrial Septum: An Unusual Cause of Paradoxical Embolism," Journal of the Saudi Heart Association: Vol. 34 : Iss. 2 , Article 3. Available at: https://doi.org/10.37616/2212-5043.1298

This Case Report is brought to you for free and open access by Journal of the Saudi Heart Association. It has been accepted for inclusion in Journal of the Saudi Heart Association by an authorized editor of Journal of the Saudi Heart Association.
Right Superior Vena Cava Connected Left Atrium with Partial Anomalous Pulmonary Venous Return and Intact Atrial Septum: An Unusual Cause of Paradoxical Embolism

Abdullah A. Al Sehly,*, Abdah Hrif, Ahmed BinSalahuddin

Abstract

A six-month-old girl presented with history of convulsion and cyanosis, was eventually found to have a right superior vena cava connected to left atrium with anomalous upper right pulmonary vein return and without atrial septal defect. Diagnosis was confirmed by cardiac magnetic resonance imaging. The patient underwent a successful cardiac surgery correction, routing SVC to right atrium and repair of right upper pulmonary vein to left atrium.

Keywords: Right superior vena cava, Paradoxical embolism, Cyanosis, Left atrium

1. Case report

A six-month-old girl referred to our hospital for further evaluation of acute onset of convulsion with persistent hypoxia. She was admitted to a local hospital after an episode of convulsion and was treated empirically with intravenous antibiotics for suspected acute meningitis. Physical examination revealed stable vital signs, apart from low oxygen saturation via pulse oximetry (75% in room air). There were no dysmorphic features and no signs of respiratory distress. Her weight was 7 kg and her height was 67 cm. Her cardiovascular and respiratory examination were unremarkable. Her neurological examination was normal apart from mild weakness of her left upper limb with mildly decreased tone and reflex. Laboratory investigation showed normal complete blood count with high hemoglobin and hematocrit. Chest X-ray showed mild cardiomegaly with clear lung fields. Her renal and liver function tests were normal. Brain magnetic resonance imaging (MRI) showed evidence of acute cerebral infarction. An echocardiography revealed atrial situs solitus, levocardia, D-loop ventricle, concordant atroventricular and ventriculoarterial connection, normal related great arteries with mild LV dilatation. There was evidence of abnormal connection of right sided superior vena cava (SVC) draining to the left atrium (LA) with anomaly of right upper pulmonary vein (RUPV) return and intact atrial septum (Fig. 1). The diagnosis was confirmed by cardiac magnetic resonance imaging (CMRI) (Fig. 2), which also revealed the unique extra abnormality of partial anomalous pulmonary venous return (PAPVR) whereas the right upper pulmonary vein joins the SVC before it enters to the LA. The rest of pulmonary veins join LA in usual fashion. No left SVC. After the patient was...
neurologically stabilized, she underwent cardiac surgery in which all the above findings were also confirmed (Image 1), the surgery was without cardiopulmonary bypass (off bump) in form of rerouting the upper part of SVC to right atrium (RA) via right atrial appendage keeping the lower part of SVC which drains the RUPV connected to LA. The patient’s O₂ saturation improved immediately post-surgery to normal and her post-operative echocardiography showed unobstructed right-sided pulmonary venous return to the LA. The right SVC drained into the RA with no flow acceleration at the junction site, the patient was discharged home in stable condition and minimal neurological sequences. Three -year follow-up showed a well-looking girl with oxygen saturation of 97% in room air with normal neurological examination. Her echocardiographic findings were similar to post-operative findings.

2. Discussion

The congenital anomalies of systemic venous connection of the heart represent a wide and heterogeneous group of malformation [1]. The development of this system begins from the symmetric and paired cardinal and subcardinal vein in the 5-week-old embryo [2]. A persistent left SVC draining to coronary sinus occurs in approximately 0.3—0.5% of healthy individual and 4.4% of patient with congenital heart disease [3]. A right sided SVC that drains into LA is a very rare congenital anomaly. Patients present with unexplained cyanosis which may be mild or severe and can present during
infancy, adolescence, or adulthood [4]. Surgical repair is needed to avoid persistent hypoxemia and prevent complications by paradoxical embolism or brain abscess [5].

Reviewing the literature, Van Praagh et al. described in 2003 three cases of anomaly drainage of right SVC into left atrium, the first patient was a 6-year-old female presented with fever, headache, and convulsion. She was diagnosed with brain abscess and her condition deteriorated rapidly and died. Her heart specimen was examined and revealed anomaly drainage of right SVC into left atrium. Second patient was an 18-h-old baby boy was admitted to neonatal intensive care due to lethargic and hypotonic with marked cyanosis. Cerebral arteriovenous (AV) fistula was diagnosed with a carotid angiogram, then he deteriorated quickly and died. Postmortem heart examination revealed two very small right upper pulmonary veins drained into the RSVC with anomaly drainage of RSVC to LA. Third patient was a 34-week gestation premature girl was presented to emergency department with cyanosis and shortness of breath. Transthoracic echocardiography revealed tetralogy of Fallot with pulmonary atresia and small atrial septal defect (ASD). The right SVC, after receiving the right upper pulmonary veins, drained exclusively into the LA. In our case report the patient was 6 months old and presented with cyanosis and convulsion, brain MRI showed acute brain infarction and echocardiography discovered to have anomaly drainage of RSVC to LA with PAPVR and intact atrial septum. Our patient underwent a successful surgical repair and discharged in stable condition. Van Praagh reviewed previously published seven cases of biatrial drainage of RSVC and 18 cases of left atrial drainage of the RSVC in viscerointestinal situs solitus [6]. Tsuneo et al. reported in 2018, a 46 old man presented with brain abscess and recurrent stroke, then was discovered to have draining of right superior vena cava (SVC) directly into the left atrium (LA), persistent left superior vena cava and atrial septal defect (ASD) [7]. In our case report, a 6-month-old girl presented with cyanosis and symptoms of stroke and had single right SVC drain into left atrium with no ASD. Recently, Hammoud et al. published one case like our case, a 17-day old newborn was discovered incidentally to have cyanosis. Echocardiography and Cardiac CT scan confirmed to have a single right SVC draining anomalously to the cranial and medial aspect of the left atrium without PAPVR or ASD. However, our case presented with cyanosis and convulsion. Additionally, there were association with abnormal right upper pulmonary vein return PAPVR [8]. The primary aim of this case report is to raise awareness about the importance of recognizing this rare anomalous systemic venous connection as one of the unusual causes.

Image 1. (A) Intraoperative photo, showing the superior vena cava (SVC) displaced distally, to show the right upper and lower veins, Note the right upper pulmonary vein (RUPV) drains directly to right SVC. The right lower pulmonary vein (RLPV) joins left atrium (LA) in normal fashion. (B) Intraoperative photo, showing SVC displaced laterally after anastomosed to right atrial appendage. Note, the lower part of SVC draining the RUPV kept connected to left atrium.
of cyanosis and paradoxical embolism with central nervous system manifestations even in the absence of atrial septum defect in different age groups. Another purpose to report this case is to pay attention of significance of using pulse oximetry test in assessment of newborns before discharge as it becomes the standard of care on detection of critical congenital heart disease in newborns [9]. Diagnosis of this congenital heart disease is set by transthoracic echocardiography, and bubble study is proved to be very helpful. The utilization of advance cardiac imaging in this rare disease is paramount important to confirm diagnosis and to delineate other associated anomalies such as pulmonary venous anomalies to avoid any surprises in operation room.

Author contribution

Conception and design of Study: AAAS, AH; Literature review: AAAS, AH; Research investigation and analysis: AAAS, AH; Data collection: AAAS, AH; Drafting of manuscript: AAAS, AH; Revising and editing the manuscript critically for important intellectual contents: AAAS, AH; Data preparation and presentation: AAAS, AH.

Source of funding

None.

Ethical approval

The study was approved by King Faisal Specialist Hospital and Research Center with Abdullah International Medical Research Center (KFSH-RC).

Conflict of interest

None.

References

[1] Mazzucco A, Bortolotti U, Stellin G, Gallucci V. Anomalies of the systemic venous return: a review. J Card Surg 1990 Jun;5(2):122–33. https://doi.org/10.1111/j.1540-8191.1990.tb00749.x. PMID: 2133830.
[2] Minniti S, Visentini S, Proacci C. Congenital anomalies of the venae cavae: embryological origin, imaging features and report of three new variants. Eur Radiol 2002 Aug;12(8):2040–55. https://doi.org/10.1007/s00330-001-1241-x. Epub 2002 Mar 19. PMID: 12136232.
[3] Teinberg I, Dubilier Jr W, Lukas DS. Persistence of left superior vena cava. Dis Chest 1953 Nov;24(5):479–88. https://doi.org/10.1016/S0012-3692(53)80048-8. PMID: 13107543.
[4] Kirsch WM, Carlsson E, Hartmann Jr AF. A case of anomalous drainage of the superior vena cava into the left atrium. J Thorac Cardiovasc Surg 1961 Apr;41:550–6. PMID: 13756294.
[5] Allen HD. Moss & adams' heart disease in infants, children, and adolescents, including the fetus and young adult. 9th ed. Philadelphia, PA: Lippincott; 2016. p. 803–4.
[6] Van Praagh S, Geva T, Lock JE, Nido PJ, Vance MS, Van Praagh R. Bialtral or left atrial drainage of the right superior vena cava: anatomic, morphogenetic, and surgical considerations–report of three new cases and literature review. Pediatr Cardiol 2003 Jul-Aug;24(4):350–63. https://doi.org/10.1007/s00246-002-0329-7. Epub 2002 Dec 4. PMID: 12457258.
[7] Nakajima T, Sakai T, Hara H. Paradoxical embolism due to right superior vena cava draining into the left atrium. Rinsho Shinkeigaku 2018 Mar 28;58(3):171–7. https://doi.org/10.5692/clinicalneurol.cn-001107. Japanese. Epub 2018 Mar 28. PMID: 29491330.
[8] Hammouda AY, Bleiweis MS, Chandran A, Steiner MB. Isolated right superior cava drainage to the left atrium: an uncommon etiology of cyanosis. World J Pediatr Congenit Heart Surg 2020 Jul;11(4):528–30. https://doi.org/10.1117/21501351.20912401. PMID: 32645773.
[9] Mahle WT, Newburger JW, Matherne GP, Smith FC, Hoke TR, Koppel R, et al. American heart association congenital heart defects committee of the council on cardiovascular nursing, and interdisciplinary council on quality of care and outcomes Research; American academy of pediatrics section on cardiology and cardiac surgery; committee on fetus and newborn. Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. Pediatrics 2009 Aug;124(2):823–36. https://doi.org/10.1542/peds.2009-1397. Epub 2009 Jul 6. PMID: 19581259.