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Isolated Right Superior Vena Cava along with Small Persistent Left Superior Vena Cava Draining into the Left Atrium-Rare Cause of Cyanosis, Think Outside the Box: A Case Report

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Abstract:-
Background: Anomalous systemic venous connection with the left atrium per se is a very rare cause of desaturation. Persistent left superior vena cava (SVC), as a variant accounts for the most common entity among all anomalous systemic connection with an incidence of 2.1% to 4.3% of the people with congenital hearts defects. The purpose of presenting this particular case is to make emphasis on evaluating the patients with isolated cyanosis meticulously. We have tried to review the previously reported cases of such anomaly under the discussion part.

Case presentation: 24-years-old young healthy lady developed symptoms of mild respiratory infection and had gone to a nearby physician Echocardiography showed that heart chambers, ejection fraction, and pulmonary artery pressure were normal. She underwent further evaluation with an HRCT and echocardiogram because of unexplained persistent desaturation. Her HRCT revealed no parenchymal lung changes which could explain desaturation. On Echocardiography, right SVC to left atrium was suspected. Chest Computed Tomography (CT) obtained after contrast material injection into the right arm vein confirmed the large right sided superior vena cava (SVC) and a small persistent left SVC draining into left atrium (LA) which explained low saturation being a significant right-to-left shunt caused by the right-sided superior vena cava (SVC) draining into the left atrium Eventually patient was referred to surgery.

Conclusions: Patient who has unexplained desaturation on clinical examination should be looked with suspicion of having anomalous systemic venous drainage. In these suspected cases of RSVC into the LA, cardiac catheterization should be performed to rule out other intracardiac lesions and to evaluate pulmonary venous drainage, and right ventricular size if there are no other definite contraindications to use contrast.

Keywords:- *Partial anomalous systemic venous drainage, Cyanosis, Left atrium.*

I. INTRODUCTION

Anomalous systemic venous connection with the left atrium per se is a very rare cause of desaturation. Persistent left superior vena cava (SVC), as a variant accounts for the most common entity among all anomalous systemic connection with an incidence of 2.1% to 4.3% of the people with congenital hearts defects [1]. With further consideration an anomalous right superior vena cava (SVC) connection to the LA is less common [2]. The purpose of presenting this particular case is to make emphasis on evaluating the patients with isolated cyanosis meticulously. We have tried to review the previously reported cases of such anomaly under the discussion part.

II. CASE HISTORY

24-years-old young healthy lady developed symptoms of mild respiratory infection and had gone to a nearby physician. While her symptoms were mild, her oxygen saturation was in low 80s. Her PCR for COVID-19 was negative and she was admitted to intensive care unit and was given supplemental oxygen. Her physical examination was unremarkable except for cyanosis. Electrocardiography (ECG) showed normal sinus rhythm. Her x-ray, lab tests and echocardiogram was normal so she underwent further evaluation with an HRCT and echocardiogram because of unexplained persistent desaturation. Her HRCT revealed no parenchymal lung changes which could explain desaturation. On Echocardiography, right SVC to left atrium was suspected. Chest Computed Tomography (CT) obtained after contrast material injection into the right arm vein confirmed the large right sided superior vena cava (SVC) and a small persistent left SVC draining into left atrium (LA) which explained low saturation being a significant right-to-left shunt caused by the right-sided superior vena cava (SVC) draining into the left atrium [Figure1].

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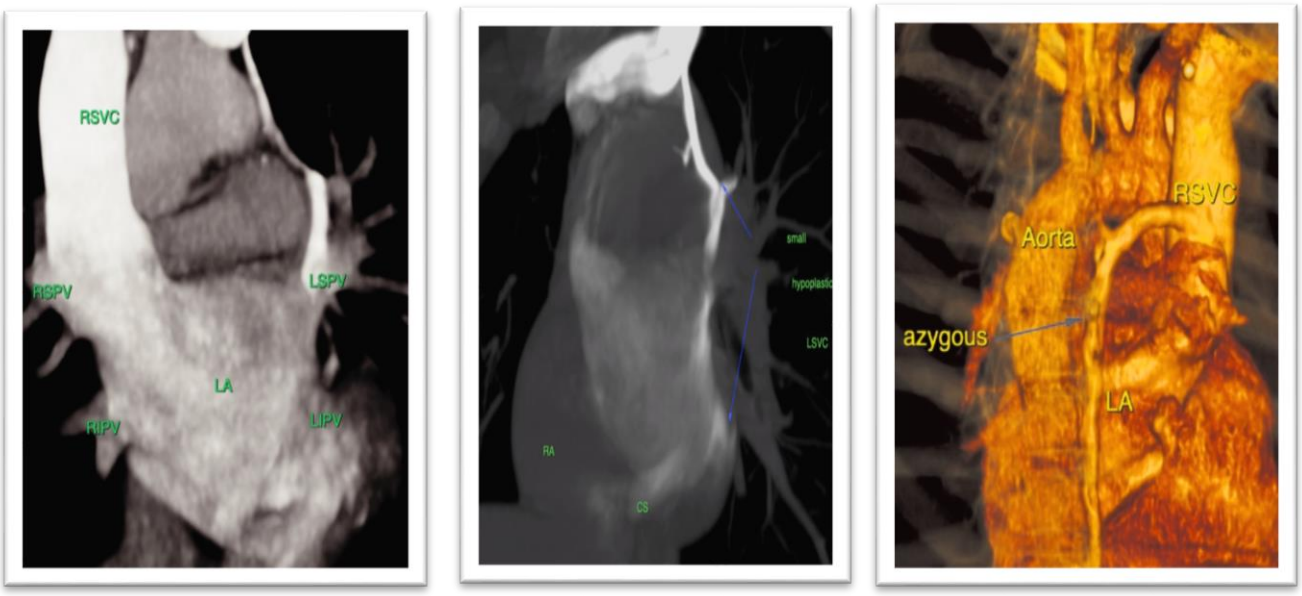

(a) (b) (c)

Fig. 1: (a,b,c) CT Images showing RSVC draning into LA, also relation with RSPV can be seen.

Cardiac catheterization was performed (Figure 2). Final diagnosis of anomalous drainage of the right superior vena cava and left small persistent superior vena cava into the left atrium with resultant right-to-left.

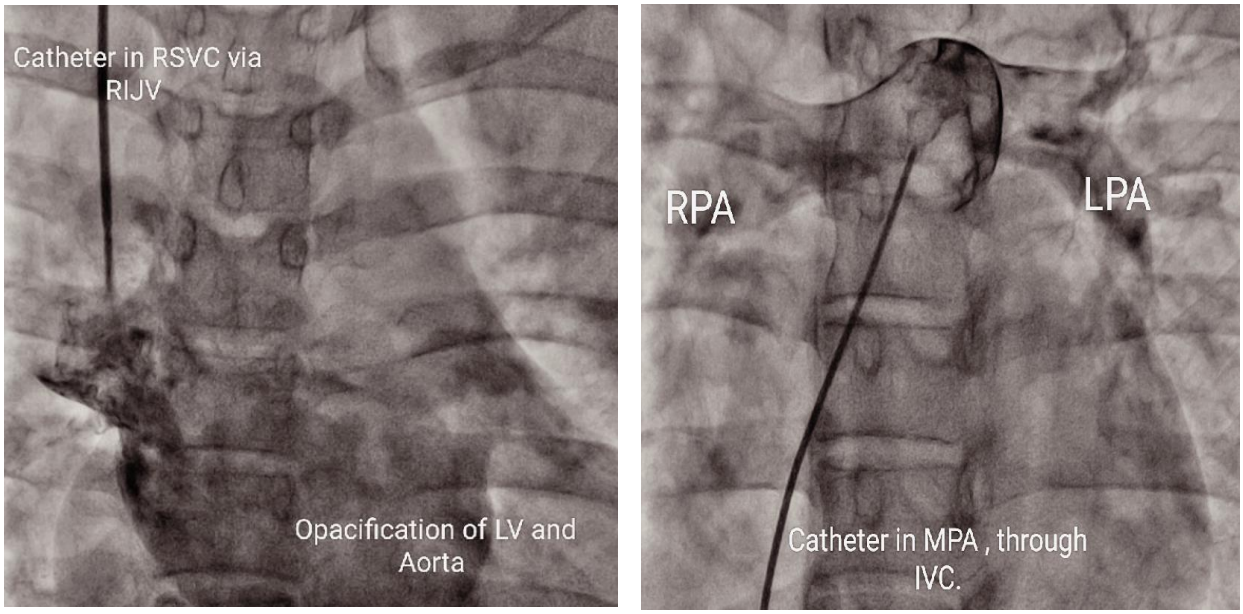

Fig. 2: Cardiac catheterization shows contrast injection into RSVC causing opacification of LA, LV and Aorta.

Shunt with normal pulmonary artery pressure was made. There was no atrial septal defect. Eventually patient was referred to surgery. Surgery was done via median sternotomy. Patient was fully heparinised. Temporary veno venous bypass was done from SVC to RA. Right superior pulmonary vein (RSPV) was found to be joining SVC and together they were going to LA. [Figure 3a] SVC was divided above the RSPV and RSPV repaired. SVC was then connected to RA. A small autologous pericardial patch was used anteriorly to augment the anastomosis. [Figure3b] Temporary venous bypass removed and SVC gradient measured on table which was found to be minimal. Post operative TEE was performed which was also satisfactory with no obstruction in SVC and RSPV.

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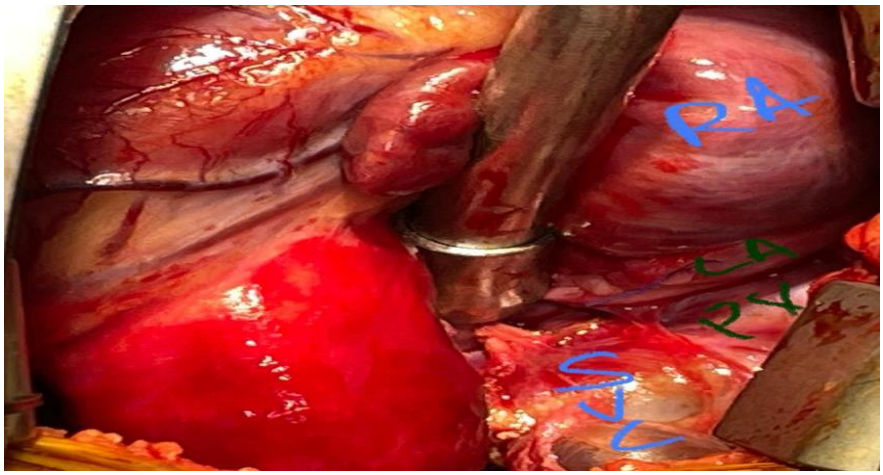

Fig. 3: Per-op image depicting the anomalous systemic connection with relation to RA and RSVP.

III. DISCUSSION

Partial anomalous systemic venous anomalies are rare but significant cause of unexplained desaturation in a structurally normal heart otherwise. Any patient who has unexplained cyanosis should be evaluated for the same. There are few similar cases reported previously who had mild hypoxemia and cyanosis [3]. First case was published in a 34-year-old hypoxemic woman who admitted to the hospital for her third childbirth [4]. Majority of these patients were suspected to have pulmonary embolism as they have presented with shortness of breath with chest pain most frequently. An abnormal venous return was suspected when radionuclide distribution in the left heart chambers and arterial system was seen injected in the upper arm [5-8]. With further detailed studies, three patients were having persistent left superior vena cava anomaly with right superior vena cava drainage into the left atrium [9-13]. In our patient, right superior pulmonary vein (RSPV) was found to be joining SVC and together they were going to LA.

IV. CONCLUSION

The conclusion of our case is that, any patient who has unexplained desaturation on clinical examination should be looked with suspicion of having anomalous systemic venous drainage. Further enquiry needs to be done to document shortness of breath, brain abscess and stroke as these are some other presenting features noted in literature. Though in our case most prominent presenting feature was cyanosis. In these suspected cases of RSVC into the LA, cardiac catheterization should be performed to rule out other intracardiac lesions and to evaluate pulmonary venous drainage, and right ventricular size if there are no other definite contraindications to use contrast. Computed tomography or magnetic resonance imaging may be required for confirmation of the side of the SVC and visualization of the anomalous right PV and for detecting other congenital anomalies. No matter how they present, treatment of choice will always be surgical correction once the diagnosis of systemic venous connection to the LA is confirmed [10]. Also, to keep in mind that intravenous infusions that use veins of the upper body should be avoided once a patient with this anomaly is recognized [14].

REFERENCES

[1.] De Leval MR, Ritter DG, McGoon DC, Danielson GK (1975) Anomalous systemic venous connection. Surgical considerations. Mayo Clin Proc 50: 599-610.

[2.] Baggett C, Skeen SJ, Gantt DS, Trotter BR, Birkemeier KL (2009) Isolated Right Superior Vena Cava Drainage into the Left Atrium Diagnosed Noninvasively in the Peripartum Period. Tex Heart Inst J 36: 611-614.

[3.] Right Superior Vena Cava Draining into the Left Atrium Songul Usalp1 *, Ali Rıza Karaci2 , Kevser Gülcihan Balcı3 , Volkan Yazıcıoglu2 , Murat Baskurt1 and Nurcan Arat Koc1 1 Department of Cardiology, Istanbul Bilim University, Turkey 2 Department of Thoracic & Cardio-vascular Surgery, Istanbul Bilim University, Turkey 3 Specialist of Cardiology, Turkiye Yuksek Ihtisas Research and Education Hospital, Turkey

[4.] Anomalous drainage of the right superior vena cava into the left atrium in a 61-year-old woman S Rosenkranz 1, A Stäblein, H J Deutsch, H W Verhoeven, E Erdmann

[5.] Nazem A, Sell JE (1993) Closed technique for repair of right superior vena cava draining to left atrium. Ann Thorac Surg 55: 1568-1570.

[6.] Pretorius PM, Gleeson FV (2004) Case 74: Right-sided Superior Vena Cava Draining into Left Atrium in a Patient with Persistent Left-sided Superior Vena Cava. Radiology 232: 730-734.

[7.] Theron A, Paule P, Metras D (2009) Right superior vena cava draining into the left atrium, associated with abnormal pulmonary vein drainage. Archives of Cardiovascular Disease 102: 249-250.

[8.] Schick EC Jr, Lekakis J, Rothendler JA, Ryan TJ (1985) Persistent left superior vena cava and right superior vena cava drainage into the left atrium without arterial hypoxemia. J Am Coll Cardiol 5: 374-378.

[9.] Leys D, Manouvrier J, Dupard T, Kassiotis P, Rey C, et al. (1986) Right superior vena cava draining into the left atrium with left superior vena cava draining into the right atrium. Br Med J 293

[10.] Schick EC Jr, Lekakis J, Rothendler JA, Ryan TJ (1985) Persistent left superior vena cava and right

superior vena cava drainage into the left atrium without arterial hypoxemia. J Am Coll Cardiol 5: 374-378.

[11.] Leys D, Manouvrier J, Dupard T, Kassiotis P, Rey C, et al. (1986) Right superior vena cava draining into the left atrium with left superior vena cava draining into the right atrium. Br Med J 293: 855

[12.] Courand PY, Girerd N, Chauveau S, Chevalier P (2013) Right superior vena cava drainage into the left atrium revealed by multiples strokes after pacemaker implantation. Heart Rhythm

[13.] Van Praagh S, Geva T, Lock JE, Nido PJ, Vance MS, et al. (2003) Biatrial 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 24: 350-363.

[14.] Isolated drainage of the superior vena cava into the left atrium in a 52-year-old man: a rare congenital malformation in the adult presenting with cyanosis, polycythemia, and an unsuccessful lung scan M D Ezekowitz, P O Alderson, B H Bulkley, P N Dwyer, L Watkins, D L Lappe, H L Greene, L C Becker