Rare congenital fistula connection between right subclavian artery and superior vena cava presenting in neonate with congestive cardiac failure

Abstract

This case report describes a congestive cardiac failure in neonate secondary to rare congenital arteriovenous fistula between the right subclavian artery and superior vena cava. The abnormality was initially discovered by echocardiography and then confirmed by CT angiography. The neonate got successful surgical repair and uneventful post surgical period. Intra-thoracic congenital arteriovenous malformations are very rare and presenting early with cardiac failure, which is challenging for diagnosis and treatment. However early identification and treatment is the most influencing factor in mortality and morbidity of these cases.

Keywords: arteriovenous malformation, subclavian artery, fistula, congestive cardiac failure

Introduction

Intra-thoracic arteriovenous malformations are uncommon. As the most reported localizations of AVM are the head (vein of Galen malformation), the abdomen (infantile hepatic hemangioma), the neck and extremities.1 Arterio venous malformation divided into congenital and acquired. The congenital forms are even more uncommon and patients present with variable symptoms that make the diagnosis more challenging.2,3 A congenital Aortocaval fistula from subclavian artery to the superior vena cava (SVC) may represent a subclass of this condition.

Case report

Our patient is full term to primigravida mother born by spontaneous vaginal delivery with good Apgar score and birth weight of 3.48 kg. Baby required no resuscitation. No risk factors for sepsis. Baby developed tachypnea at 2 hours of life, so shifted to Special care Baby Unit (SCBU). Examination revealed bruit over right subclavian area.

Echocardiography was done on day one of life and showed:

a. Situs solitus, levocardia.

b. The SVC was dilated and had high pulsatile flow.

c. The RA and right ventricle (RV) were moderately dilated.

d. Milder Poor ventricular function, ejection fraction (EF) 51%.

e. Mean pulmonary artery (MPA) was dilated (9.2mm) and branches are normal.

f. Pulmonary veins all present with normal drainage to left atrium (LA).

g. There was a 3 mm patent ductus arteriosus (PDA) with mainly Rt to LT flow.

h. Arch was left and the first branch was dilated. There was significant diastolic runoff in the transverse and descending aorta, no narrowing or sign of CoA.

Impression: Extra cardiac arteriovenous shunt, Right subclavian artery (SCA) to SVC

Baby started on anti failure medications. The baby deteriorated on day 2 of life, so incubated. Echo repeated with same findings plus newly developed pulmonary hypertension (Figure 1) (Figure 2).

Figure 1 Short axis view Echocardiography for the AV connection between the dilated SCA to Dilated SVC. Baby started on anti failure medications. The baby deteriorated on day 2 of life, so incubated. Echo repeated with same findings plus newly developed pulmonary hypertension.

CT Angiography showed:

a. Anomalous vessels connecting proximal right subclavian artery with superior vena cava, representing arteriovenous fistula.

b. Cardiomegaly and right sided heart strains likely related to arteriovenous shunting.
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Discussion

Aortocaval fistulas are rare cause of left-to-right-shunt resulting in neonatal congestive heart failure. The vessels initially develop from the mesenchyma in an undifferentiated form till give fully developed veins and arteries. So, they develop from same primordial network of vessels. Congenital arteriovenous fistulas may be the result, if these early connections of both arteries and veins persist. Hereby congenital AVM is a mal-development of blood vessels, with preservation of one or more primitive direct communications between arterial and venous channels. Histological, the fistula connection shows hyperplasia of elastic fibers of the arterialized vein, ranging from 41 to 82%. There are few cases in literature similar to our case, right subclavian artery to SVC fistula reported in 1977 by Arkell and Lawson, in 2011 case by Balakrishnan et al., and another case by Awasthy et al. Three case reported by Gutierrez et al., two of them showed a fistula between left subclavian artery-to-innominate vein and draining into markedly dilated superior vena cava. Saire et al. In 1983 reported a case of arterio-venous fistula between the subclavian artery and in nominate vein presented with heart failure in neonatal period. In 2000 Dogan et al., & Tatum et al., in 2006 reported congestive heart failure due to subclavian artery to subclavian vein fistula. In 2016, Sami Jabari & Robert Cesnjevar reported a case of fistulatous connection between Brachiocephalic trunk and superior vena cava. The echo cardio graphy is simple fast screen tools for cardiac abnormality and first alert to such entity. The confirmation by using 64 slice CT scan was hand-to-hand a great support that confirm the diagnosis which facilitate the transfer of patient to the appropriate institution to get the proper treatment.

Conclusion

The early presentation of heart failure in neonatal period necessitate extensive work up excluding all arteriovenous malformation in heart, head, chest and abdomen. This case highlights a rare congenital arteriovenous fistula connection between subclavian artery and superior vena cava. Echocardiography is important non invasive tool in diagnosing and raising suspicion of congenital Aortocaval arteriovenous fistula. CT angiography was complementary tool to diagnose the malformation.

References


Citation: Al-Majrafi AS, Kalbani NKA, Ambusaidi F. Rare congenital fistula connection between right subclavian artery and superior vena cava presenting in neonate with congestive cardiac failure. *J Pediatr Neonatal Care*. 2018;8(3):148–149. DOI: 10.15406/jpnc.2018.08.00328