A rare case of congenital thoracic arteriovenous fistula between the brachiocephalic trunk and the superior vena cava resulting in heart failure

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A B S T R A C T

We present a case of a full-term 13 days old female baby administered to the PICU with extreme tachypnea with suspicion of a huge left-to-right-shunt at vascular or cardiac level. Echocardiography revealed an open arterial duct and a huge arterio-venous fistula from the proximal right subclavian artery directly into the superior vena cava. The fistula was cross-clamped, resected and both vessel-stumps were oversewn. Histopathologically the fistula presented either as an artery of the elastic type or an arterialized vein, which is a typical finding in fistulas.

The patient was extubated shortly after repair and referred home five days after surgery. To the best of our knowledge, this is first report of a direct fistulous connection between the brachiocephalic trunk (proximal to the right subclavian artery) and the SVC with congestive heart failure and ASD.

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Aortocaval fistulas are a rare cause of a left-to-right-shunt. Most of the fistulas are located intracerebral, cervical or intraabdominal. Intrathoracic aortocaval fistulas are very rare of them congenital fistulous connections are extremely rare. A congenital aortocaval fistula from the brachiocephalic trunk to the superior vena cava (SVC) may represent a subclass of this condition. In the following such a case is presented. The fistula was successfully closed.

1. Case

Pediatric intensive care unit admission of a full-term 13 days old female baby (41st week of gestation) with extreme tachypnea. Echocardiography revealed a massively enlarged right ventricle and right atrium. Patient was transferred to our tertiary care center immediately with suspicion of a huge left-to-right-shunt at vascular or cardiac level. Echocardiography in detail showed an open arterial duct and a huge arterio-venous fistula from the proximal right subclavian artery directly into the superior vena cava. As to our knowledge the patient had not had any intervention such as line placement prior to presentation which could have caused a traumatic fistula. Chest-X-Ray revealed huge enlargement of the overshunted heart. An interventional attempt during pre-operative cardiac catheterization to close the duct and the huge fistula was not undertaken, because placement of a device would have caused either obstruction or occlusion of the right subclavian artery or the superior caval vein. Thus the patient was referred for surgical repair. The chest was opened via median sternotomy and the arterial duct was closed routinely. The fistula presented either as an artery of the elastic type or an arterialized vein, which is a typical finding in fistulas.
2. Histology

The wall of the fistula was thickened and composed mainly of hyperplastic elastic tissue, appearing either as an artery of the elastic type or an arterialized vein, which is a typical finding in fistulas [1,2]. There were no signs for infection or malignancies (Fig. 1).

3. Embryology

In the mesenchyma the vessels initially develop in an undifferentiated form and then grow and regress until completion of development. Hereby veins and arteries derive from the same primordial network of vessels. If these early connections of both arteries and veins persist, congenital arteriovenous fistulas may be the result [3,4].

4. Discussion

Arteriovenous fistulas are rare and most reported cases may be classified as acquired i.e. secondary to trauma or rupture of a dissecting aneurysm in Marfan syndrome [5]. Congenital arteriovenous malformations are even rarer, but an important cause of neonatal heart failure. Common localizations are the head (i.e. lesions of the vein of Galen), the neck, the extremities and the abdomen (hepatic hemangioendothelioma, kidneys) [6–8]. Intra-thoracic arteriovenous malformations, however, are far less common.

Among them, the commonest forms include those that arise from the aortic root or ascending aorta and that are associated with rupture of the sinus of valsalva or a dissecting aneurysm and coronary fistula. These arteriovenous fistulas commonly drain into the right cardiac chambers and pulmonary arteries [5,9].

Nevertheless, these fistulas can originate from any part of the aorta, the coronary arteries and the intercostal or mammary arteries. They drain into the innominate vein, the azygote vein, the superior vena cava, the right atrium and pulmonary circulation [10–19]. Soer et al. [20] described an unusual case of a congenital systemic arteriovenous fistula arising from the descending aorta and draining separately into the SVC, azygos vein, and innominate vein.

Fistulous connections from the subclavian artery into systemic veins however have been reported only eight times before.

Walker et al. [21] described a case of a massive arteriovenous fistula between the left subclavian artery and innominate vein, which was revealed after the death of the child by necropsy.

Arkell and Lawson [22] described another case of an arteriovenous fistula from the right subclavian artery into the SVC. Histology of the excised vessel demonstrated two vascular channels, consisting of thick-walled muscle with increased subintimal connective tissue. No evidence of atheroma or inflammatory endarteritis was found. In 1983 [23] a case of subclavian artery to innominate vein fistula associated with congestive heart failure in a newborn was presented by Sapire et al. Gutierrez et al. [11] reported three cases, two of them with fistulous connections from subclavian arteries to the innominate vein and azygous vein respectively. In 1989, Wong et al. reported congenital myxedema and a fistulous connection from an aberrant right subclavian artery to the SVC [24]. In 2010 Awasthy et al. described the case of child with a fistulous connection of the right subclavian artery to SVC and its nonsurgical management [25]. The latest report of a subclavian artery to SVC fistula resulted in neonatal heart failure and was presented by Balakrishnan et al. [26].

To the best of our knowledge, this is first report of a direct fistulous connection between the brachiocephalic trunk (proximal to the right subclavian artery) and the SVC with congestive heart failure and ASD.

Conflict of interest

The authors have declared that no conflict of interest exists.

References


