A Rare Anatomic Variation: A Combination of Anomalous Origin of the Right Subclavian Artery from the Main Pulmonary Artery, Ventricular Septal Defect, and Aortic Coarctation

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ABSTRACT

Background: We present a rare case of an anomalous origin of the right subclavian artery (SA) from the pulmonary artery (PA) associated with ventricular septal defect (VSD) and aortic coarctation.

Case Report: Critical aortic coarctation and VSD were diagnosed in a neonate, and coarctation angioplasty was successfully performed. Severe cardiac failure developed after this procedure, however, and closure of the VSD was planned.

Results: The anomalous SA was diagnosed during the operation when the cardiopulmonary bypass was initiated. As the pulmonary blood flow decreased, a difference in pulse pressures between the right radial artery and the ascending aorta was noticed to be due to the subclavian steal phenomenon. The difference decreased from 60 mm Hg to 25 mm Hg following ligation of the SA at its origin from the PA. The patient was discharged on the eighth postoperative day without any problems.

Conclusion: An abnormal origin of the right SA from the main PA associated with VSD and aortic coarctation is a unique combination that, if unnoticed preoperatively, may cause difficulties during the operation.

INTRODUCTION

Cases of a subclavian artery (SA) abnormally originating from the pulmonary artery (PA) have occasionally been reported. Most of these are abnormalities of the left SA associated with a right arch abnormality. An abnormal origin of the right SA from the PA is scarce. In this report, we present a case of anomalous origin of the right SA from the PA associated with ventricular septal defect (VSD) and aortic coarctation.

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Figure 1. The preoperative angiogram reevaluated retrospectively revealed that the right subclavian artery (RSA) was filled in the late phase by the vertebral blood flow. RVA indicates right vertebral artery; LVA, left vertebral artery; LSA, left subclavian artery; RSA-PA, right subclavian artery from the pulmonary artery; PA, pulmonary artery.
to the aortic pressure. Furthermore, the 2-dimensional images from the suprasternal notch were of low resolution, precluding a diagnosis of the right SA abnormally originating from the PA.

Cardiac catheterization revealed a PA pressure of 86/61 mm Hg. The infant underwent balloon coarctation angioplasty, and the gradient decreased to 8 mm Hg. The abnormal anatomy of the right SA could not be seen in the preoperative assessment; however, in a retrospective evaluation, the right SA was observed late following aortic injection as it was filled by the vertebral blood flow (Figure 1). In the preoperative period, the patient had no clinical findings of the subclavian steal phenomenon because of the increased PA pressure and the intraaortic shunt.

Operation

The patent ductus arteriosus was ligated just before starting cardiopulmonary bypass (CPB). With the patient under moderate hypothermia, CPB was begun after cross-clamping. The heart was arrested with blood cardioplegic solution. After cardiac arrest, the arterial pressure decreased and was normalized by increasing the pump flow. The decrease in arterial pressure was caused by the decreased pulmonary blood flow and pressure, causing steal from the right SA to the PA. Invasive arterial monitoring revealed a gradient of 60 mm Hg between the ascending aorta and the right radial artery. The VSD was repaired with a Dacron graft during 43 minutes of cross-clamping. After the patient was removed from the pump, the abnormal right SA originating from the main PA bifurcation was determined, and it was ligated at its origin from the PA. In the setting of a complex procedure in a sick newborn who requires a longer operation, we prefer simple ligation. At this point, the blood was supplied to the right SA by collaterals from the vertebral artery. This supply produced a decrease in the pressure gradient from 60 mm Hg to 25 mm Hg (Figure 2).

RESULTS

Cyanosis or ischemic signs were not recorded on the right arm in the postoperative period. The oxygen-saturation values were the same in both arms. Cerebral symptoms suggesting an SA steal phenomenon were not observed. The patient was discharged from the hospital on the eighth day of the operation.

One month after the operation, the blood pressure on the right arm was 78/43 mm Hg and was 75/52 mm Hg on the left arm. Neither cyanosis nor circulation failure was detected. On the second month of follow-up, the patient underwent catheterization for recoarctation. The angiogram revealed that the connection between the right SA and the main PA was obliterated, that the circulation of the arm was provided by collateral arteries, and that there was no reversal of flow from the vertebral artery to the right SA (Figures 3 and 4).
DISCUSSION

The condition in which the continuity of the aortic arch and the SA is interrupted and the connection between the SA and the homolateral PA is provided by a patent or closed ductus arteriosus has been described as “the isolation of SA.” This definition was first used in 1963 by Stewart et al [1963]. SA isolation has always been observed on the opposite side of the aortic arch. Although the combination of right aortic arch and left SA isolation has been reported in the literature, the present malformation of right SA isolation and the left arch is very rare. Edwards explained the embryonic formation of an isolated right SA as the regression of the right arch in 2 places. These places are (1) at the dorsal right SA and the conjunction of the ductus and the arch, and (2) between the right common carotid and the right SA.

Isolated SA presents with different clinical findings depending on the accompanying cardiac lesions. In the diagnosis, evaluation of the echocardiographic, computed tomographic (CT), magnetic resonance (MR) imaging, and angiographic examinations may be needed, depending on the accompanying cardiac lesions [McElhinney 1998]. CT and MR angiography are noninvasive methods that are important in diagnosis [Marin 2008]. Nevertheless, this rare malformation is commonly diagnosed during intraoperative dissection [Mulay 1997]. In that case, the subclavian steal syndrome is of critical importance in patients who are to undergo a systemic-pulmonary shunt operation [Hofbeck 1991; Baudet 1992; Mosieri 2004].

The blood flow in isolated SA is generally derived from the collateral circulation from the opposite SA. When the isolated SA is perfused from the vertebral artery, it may cause a subclavian steal phenomenon and produce cerebral symptoms, but these symptoms are uncommon in young patients whose PA resistance is increased [Hofbeck 1991; Baudet 1992]. Blood pressure and pulse changes and permanent or persistent cyanosis of the arm may be the clinical findings associated with this anomaly. When there is a right-to-left shunt due to increased PA pressure and right heart pressure, cyanosis may be seen on the affected extremity. It may be asymptomatic in cases in which the collateral circulation is efficient, whereas ischemic signs leading to atrophy may be observed in cases of deficient circulation [Hofbeck 1991; Baudet 1992; Mulay 1997; McElhinney 1998; Mosieri 2004].

The options for surgical correction are obliteration of the pulmonary connection by ligation of the vessel and reimplantation of the SA to the aorta. It has been doubted that artery ligation without implantation to the aorta may cause subclavian steal syndrome due to insufficient collateral circulation. Despite these risks, ligation is the most commonly used surgical method in patients who require long operations, mostly because of accompanying cardiac lesions, and is applied easily and quickly [Mulay 1997; McElhinney 1998; Mosieri 2004].

A right SA originating from the PA is a very uncommon malformation that may or lead to different clinical symptoms, such as cyanosis, ischemia, atrophy, left-to-right shunt causing pulmonary congestion, and cerebral symptoms due to subclavian steal syndrome. Depending on the presence of subclavian steal syndrome, depending on the presence of other cardiac lesions [Hofbeck 1991; Mulay 1997; Mosieri 2004]. For the diagnosis, CT and MR angiography, as well as conventional angiography, are also beneficial, although some cases may be noticed during operation with attentive surgical dissection [Hofbeck 1991; Mosieri 2004]. Ligation with or without aortic reimplantation techniques is used in surgical treatment.

REFERENCES


