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Congenital Subclavian Aortic Steal Syndrome: Hypoplastic Aortic Arch With Severe Coarctation of the Aorta of Unusual Location

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An unusual case of subclavian-aortic steal syndrome in a pediatric patient with hypoplastic aortic arch with coarctation of the aorta of unusual location is described. Aortography showed hypoplasia of the arcus aorta and severe coarctation proximal to the left subclavian artery associated with an aneurysm formation on the isthmus and descending aorta. The enlargement of the arcus aorta was accomplished by prosthetic patch aortoplasty extending from the ascending to descending aorta via median sternotomy using cardiopulmonary bypass and moderate hypothermia. Postoperatively, the patient is doing well with equalized blood pressure.

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The occurrence of subclavian aortic steal syndrome in the pediatric age group is rare. It is produced as a result of reversal of blood flow in the vertebral artery because of the occlusive lesions of the aortic arch and isthmus. It has been reported in patients with interrupted aortic arch and hypoplastic aortic arch with or without aortic coarctation [1, 2]. In this study, we present an unusual case of subclavian aortic steal syndrome in a child with a hypoplastic aortic arch and an unusually located coarctation associated with aneurysm formation in isthmus and descending aorta.

A 5-year-old girl was admitted to the hospital for an episode of syncope. Before admission, she had com-



Fig 1. Aortic arch aortography, at early phase. Hypoplastic aortic arch beyond the innominate artery and elongated left carotid artery before the coarctation are observed.

plained of headache with exertion. Physical examination results revealed a well-developed girl in no distress. On admission, pulses in the left radial, brachial, and both femoral and distal arteries were absent. Her blood pressure in the left arm was 80/50 mm Hg and in the right arm was 125/80 mm Hg. Blood pressure in both legs was found to be 80 mm Hg. On cardiac auscultation, systolic murmur was noted at the second intercostal area. A bruit was heard over the left supraclavicular region and the left neck. In addition, thrill was palpated above the sternal notch.

Electrocardiogram showed normal findings for her age. Chest roentgenogram revealed a normal-sized heart with a prominent aortic knob. The echocardiographic examination verified the aortic arch hypoplasia and coarctation. No associated lesions including ductus arteriosus and coarctation at the typical location or any other major cardiac abnormality were found. Angiogram showed severe hypoplasia of aortic arch beyond the innominate artery. The left carotid artery was elongated and tortuous. The left subclavian artery and the descending aorta could not be opacified at the same time with the proximal aortic arch, because the distal aortic arch was very narrow and occluded by the catheter (Fig 1). A severe coarctation before the left subclavian artery was noted, and the left subclavian artery branched from the dilated descending aorta. Retrograde flow, filling the proximal left subclavian artery and the descending aorta via the left vertebral artery, was noted (Fig 2).

The operation was performed through a median sternotomy using cardiopulmonary bypass and moderate hypothermia without circulatory arrest. The entire aortic arch including all its branches were widely dissected. The left carotid artery and the left subclavian artery were temporarily occluded. Curved vascular clamps were ap-

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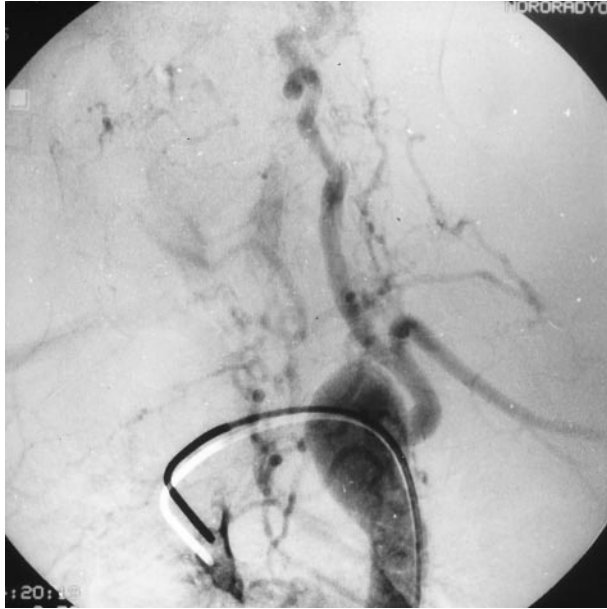


Fig 2. Retrograde filling of the dilated descending aorta and the left subclavian artery via the left vertebral artery.

plied to the arcus aorta beyond the innominate artery and descending aorta after the origin of the subclavian artery. The concavity of the arcus aorta was incised proximally from the descending aorta to the ascending aorta. A standard polytetrafluoroethylene (PTFE) patch was sutured with continuous 6-0 polytetrafluoroethylene sutures, starting at the descending aorta and carried through the ascending aorta. Intraoperative measurements revealed no pressure gradients between the ascending and descending aorta. No postoperative neurologic complications were encountered.

The postoperative course was uneventful. Two-dimensional echocardiographic studies confirmed the correction of arch hypoplasia. No pressure gradient was found between the arms and legs at the most recent examination of the 3-year follow-up.

Comment

Congenital subclavian steal syndrome resulting from disease of the subclavian artery proximal to the origin of the vertebral artery and various anomalies of the aortic arch are well recognized [3-5]. When the subclavian artery arises distal to the narrowest segment of distal aortic arch, blood flow to the lower extremity is supplied via vertebral and subclavian arteries from the basillary system and the circle of Willis causing the subclavian aortic steal syndrome and symptoms of cerebral ischemia. The vertebral artery, which is the first branch of the subclavian artery, leads to the reversal of blood flow from the right to the left subclavian artery. It has been reported in patients with interrupted aortic arch without ductus arteriosus [1, 2].

In our case, there was a hypoplastic aortic arch and a severe coarctation proximal to the left subclavian artery and aneurysm formation on the descending aorta be-

cause of jet flow through the coarctation. The well-developed left vertebral and subclavian artery supplied the left arm, and lower extremities resulted in subclavian aortic steal. Although it is known that during childhood no cerebral symptoms usually appear, our case and a few reports in the literature provide evidence that cerebral symptoms may present during childhood, even in the newborn period [4, 6].

The type of surgical repair of complex aortic arch hypoplasia is chosen according to angiographic and operative findings. In similar cases, extended end-to-end repair or patch aortoplasty has been used successfully. However, prosthetic patch aortoplasty (which is widely used in repair of aortic coarctation and aortic arch hypoplasia) offers the less radical means of reconstruction of the transverse arch and thus reduced operative risk.

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Tracheobronchial Glomus Tumor

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Glomus tumors are uncommon. A review of the literature for tracheobronchial glomus tumors revealed 13 tracheal glomus tumors. The diagnosis may be elusive and so the true incidence of tracheobronchial glomus tumors may be greater than that reported. Three of the 14 glomus tumors were initially believed to be carcinoid. Glomus tumors should be included in the differential diagnosis of tracheobronchial tumors.

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