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Case Report / Olgu Sunumu

Staged surgical treatment in an infant with left hemitruncus

Sol hemitrunkuslu bir infantta aşamalı cerrahi tedavi

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Left hemitruncus (aortic anomalous origin of the left pulmonary artery) is relatively rare than the right hemitruncus. Early diagnosis is critical to prevent early occurrence of pulmonary vascular obstructive disease in the left pulmonary bed in patients with tetralogy of Fallot presenting with left hemitruncus arteriosus. To the best of our knowledge, there are only five surviving cases with left hemitruncus and tetralogy of Fallot who were operated in the first decade of life in the literature. Any pediatric case undergoing staged surgery with full recovery has not been reported yet in the literature. In this article, we report a one-year-old girl with tetralogy of Fallot and left hemitruncus with a late diagnosis who underwent staged correction surgery with palliative left pulmonary artery banding followed by total repair.

Keywords: Child; congenital heart disease; hemitruncus; pulmonary hypertension; right arcus aorta; selective pulmonary banding; tetralogy of Fallot.

Sol hemitrunkus (sol pulmoner arterin aort kökeninden ayrılma anomalisi) sağ hemitrunkusa göre daha nadir görülür. Fallot tetralojisi ile sol hemitrunkus birlikteliğinde sol pulmoner yatakta erken gelişebilecek pulmoner vasküler obstrüktif hastalığı önlemek için erken tanı çok önemlidir. Bildiğimiz kadarıyla, literatürde, Fallot tetralojisi ve sol hemitrunkus birlikteliğinde ilk dekatta yapılan ve yaşayan beş sol hemitrunkuslu olgu mevcuttur. Aşamalı cerrahi yapılarak tam düzeltme sağlanan herhangi bir pediatrik olgu literatürde henüz bildirilmemiştir. Bu yazıda, palyatif sol pulmoner band ameliyatının ardından total tamir ile aşamalı düzeltme cerrahisi yapılan geç tanı konulmuş Fallot tetralojili ve sol hemitrunkuslu bir yaşında kız çocuğu sunuldu.

Anahtar sözcükler: Çocuk; doğuştan kalp hastalığı; hemitrunkus; pulmoner hipertansiyon; sağ arkus aort; selektif pulmoner banding; Fallot tetralojisi.

Hemitruncus, the anomalous origin of one of the pulmonary arteries from the ascending aorta, is a rare anomaly, and left hemitruncus (originating from the left pulmonary artery) is less common than right hemitruncus, which is generally associated with tetralogy of Fallot (TOF).[1] In untreated cases, the survival rate is very low because the ipsilateral pulmonary bed is vulnerable to the early onset of pulmonary vascular obstructive disease; hence, identifying hemitruncus is essential in order to prevent horrendous outcomes.[2-4] In this article, we present a case of left hemitruncus associated with TOF that was diagnosed in a one-year-old patient via an echocardiographic examination. To our knowledge, no other case like ours, in which the patient underwent staged surgical treatment, has been reported previously in the literature. In addition, our patient is also the sixth to survive after successful surgical repair.

CASE REPORT

A one-year-old girl was referred to our center because of suspected congenital heart disease and a finding of cyanosis. A physical examination revealed central cyanosis, a pulse rate of 150 beats per minute (bpm),

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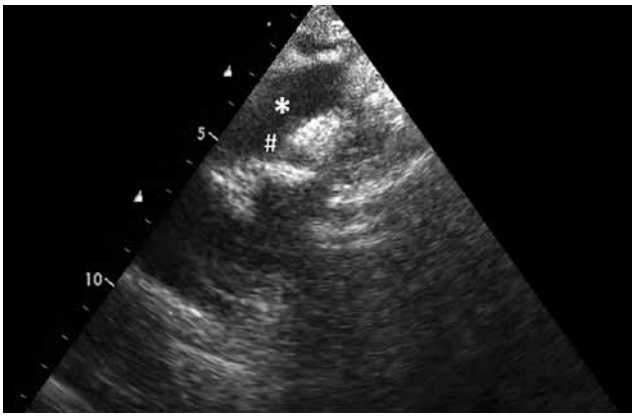

Figure 1. Transthoracic echocardiographic (TTE) image showing the suprasternal view of the abnormal vascular structure, which measured 8 mm, originating from the ascending aorta and directed toward the left lung. * Ascending aorta; # Left pulmonary artery.

and a resting systemic oxygen saturation (OS) of 90%. In addition, the right ventricular beat was hyperdynamic on palpation, and a grade 3/6 continuous murmur was heard over the pulmonary area on auscultation. An electrocardiogram showed normal sinus rhythm, right axis deviation, and right ventricular hypertrophy, and a chest X-ray detected increased pulmonary density in the left lung and decreased pulmonary vasculature in the right lung. The patient had no familial history of any congenital heart disease. In addition, her prenatal history was unremarkable which eliminated the possibility of any exposure to teratogenic agents.

The two dimensional (2D) transthoracic echocardiogram (TTE) showed normal visceroatrial situs, levocardia, TOF with a large misaligned ventricular septal defect (VSD), right ventricular hypertrophy, an overriding aorta, infundibular stenosis, a right aortic arch, a secundum atrial septal defect (ASD), and the absence of patent ductus arteriosus (PDA). The main and right pulmonary arteries were also mildly hypoplastic (6 mm). In the suprasternal view, the left pulmonary artery was seen arising anomalously from the left lateral aspect of the ascending aorta rather than the main pulmonary artery, which indicated left hemitruncus (Figure 1).

The right and left ventricular systolic pressures were equal in the cardiac catheterization. A right ventriculogram showed that the right pulmonary artery continued as the sole branch of the main pulmonary trunk, and a left ventriculogram and an aortogram (left anterior oblique projection) found that the left pulmonary artery was arising from the ascending aorta and that a right-sided aortic arch was present (Figure 2a, b). The saturation values in the blood gas before adenosine admission were 72% in the systemic veins, 87% in the left pulmonary artery, and 86% in the aorta, and the mean pressure values were 45 mmHg in the left pulmonary artery and 39 mmHg in the aorta. After administering adenosine, the saturation values were 74% in the systemic veins, 80% in the left pulmonary artery, and 88% in the aorta. In addition, the mean pressure values were 41 mmHg in the left pulmonary artery and 36 mmHg in the aorta. Furthermore, before giving the patient adenosine, the pulmonary-systemic flow ratio (Qp/Qs) was 1.2, the pulmonary vascular resistance (PVR) was 11.3 Wood Unite (WU), the systemic vascular resistance (SVR) was 9 WU, and the PVR/SVR was 1.23, whereas after the administration of this drug, the Qp/Qs was 1.04, the PVR was 8.8 WU, the SVR was 7.5 WU, and the PVR/SVR was 1.17. Therefore, the left pulmonary

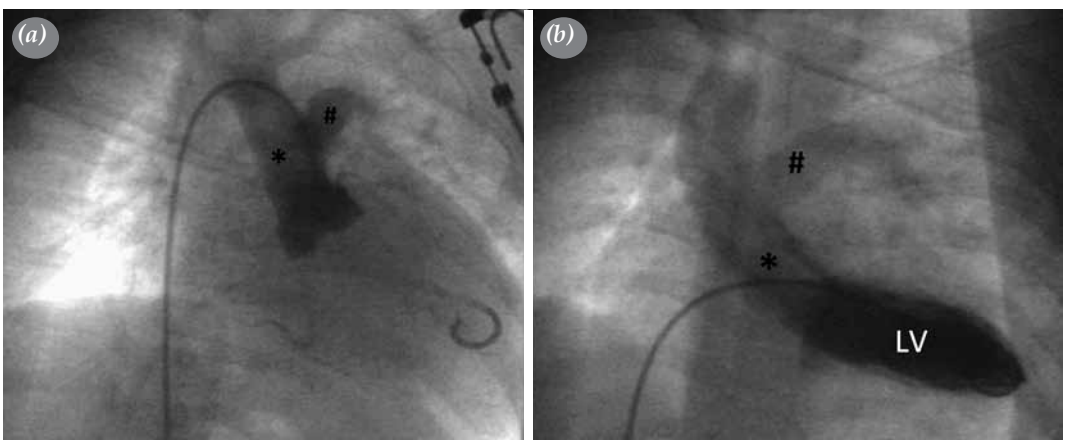

Figure 2. (a) A ventriculogram and (b) an aortogram of the left oblique region showing the abnormal vascular structure in the left pulmonary artery when the patient was 14 months old. This led to the diagnosis of the left hemitruncus. * Ascending aorta; # Left pulmonary artery; LV: Left ventricle.

vascular bed was determined to be non-reactive since the left pulmonary artery systolic pressure did not decrease significantly.

Selective pulmonary banding was then planned, and the patient was given bosentan, a pulmonary antihypertensive medication. The left pulmonary artery banding was performed (gradient of 40 mmHg) when she was 14 months old, and cardiac catheterization was repeated at the 30th month. The mean pressure value at the left pulmonary artery distal to the banding level was then measured at 52 mmHg while it was 88 mmHg at the aorta. When the patient was three years old, the correction process was finally completed.

The patient underwent an angiocardiography one year later (Figure 3), and the saturation values in the blood gas were 76% in the systemic veins, 86% in the left pulmonary artery, and 98% in the aorta. Additionally, the mean pressure values were 28 mmHg in the left pulmonary artery, 65 mmHg in the aorta, 6 mmHg in the right atrium, and 9 mmHg in the left atrium. Furthermore, the Qp/Qs was 1.7, the PVR was 2.69, the SVR was 15.37, and the PVR/SVR ratios was 0.175.

In the first operation, the left pulmonary artery arising from the ascending aorta was approached via a left minithoracotomy. Pulmonary banding of the left pulmonary artery was performed on the beating heart, which was adjusted to leave a systolic pressure of 30 mmHg at the distal portion of the band. Postoperative echocardiography revealed a 40 mmHg pressure gradient at the level of the pulmonary banding, and the patient was discharged on the fourth postoperative day.

In the second surgery, the heart was approached via a median sternotomy, and the procedure was performed using cardiopulmonary bypass (CPB) under moderate systemic hypothermia with cold cardioplegic arrest. The VSD was repaired with a Dacron patch, and the right ventricle outlet tract (RVOT) was dissected. The left pulmonary artery was also separated from

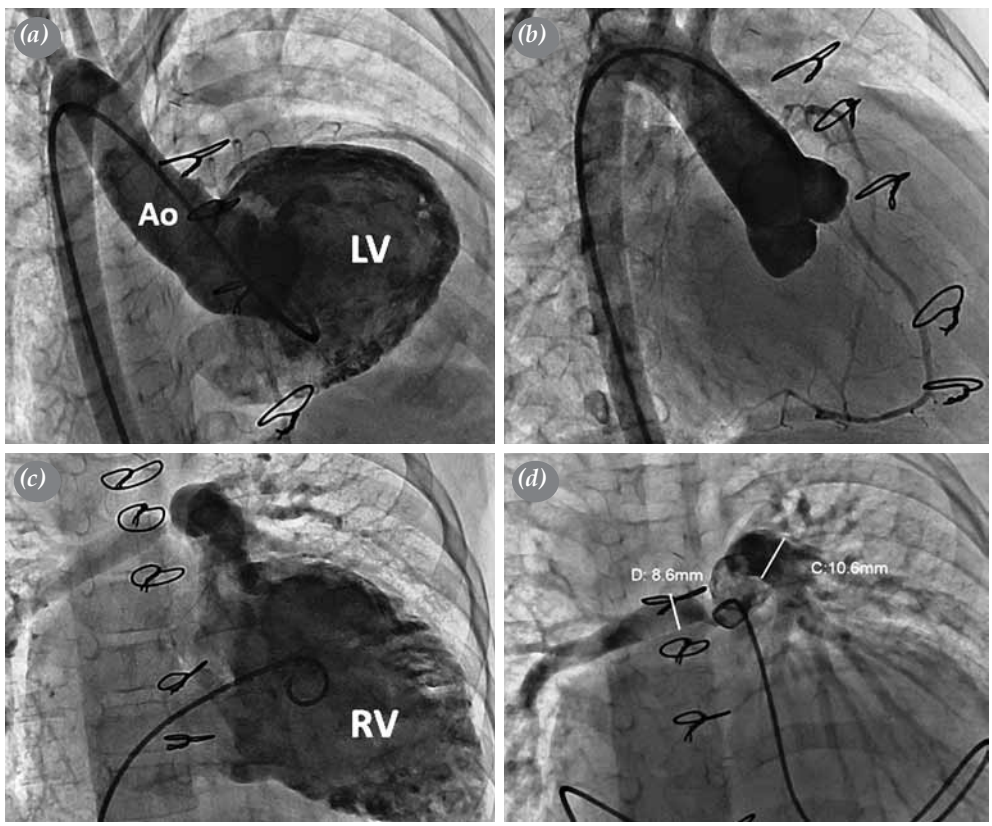

Figure 3. Angiography showing no significant hemodynamic dysfunction after the second operation. The apparent improvement was revealed in the mean pressure of the left pulmonary artery, and the mild gradient (12 mmHg) was measured in the valved conduit. The left pulmonary artery measured 8.6 mm while the right pulmonary artery measured 10.6 mm. LV: Left ventricle; Ao: Ascending aorta; RV: Right ventricle.

the aorta and unifocalization was performed on the dilated right pulmonary artery. In addition, the Contegra® 16 mm valved conduit (Medtronic, Inc., Minneapolis, MN, USA) was anastomosed between the right ventricle and pulmonary artery, and milrinone was administered for three days postoperatively. The convalescent period was uneventful, and the patient was discharged on the seventh postoperative day without any complications. An echocardiographic examination showed no gradient through the RVOT or the branches of the pulmonary artery. The patient is now three years old and asymptomatic. Moreover, recent echocardiography has detected no important hemodynamic problems.

DISCUSSION

Hemitruncus, the anomalous origin of one of the branch pulmonary arteries from the ascending aorta with two normal semi-lunar valves, is a rare entity as there have only been several small case series reported.[1-4] Compared with right hemitruncus, left hemitruncus is seldom seen, and to date, only 24 cases have been reported, with having been primarily associated with TOF.[3] Although Changela et al.[4] reported on 2,235 TOF patients, only eight were found to have right hemitruncus, and none had left hemitruncus in their study.

To diagnosis this anomaly, electrocardiography and chest X-rays are important, but echocardiography is essential because the abnormal vessel pattern can be seen on 2D and Doppler examination in the subcostal and suprasternal view. It is also important to distinguish this anomaly from discontinuous pulmonary arteries in which the blood supply to the left pulmonary artery originates from the major aortopulmonary collaterals or the ductus arteriosus.[1-3]

The primary treatment for left hemitruncus is surgical correction. Early repair of this lesion is important to improve the survival rate, which has been reported at less than 30% if left untreated.[4] New treatment modalities can be used with the improvement of developing technology and conduits, but the preferred option is primarily correction.[1-6] This was planned for our case at admission, but the patient had isolated left-sided severe pulmonary hypertension. Therefore, cardiac catheterization and a pulmonary reactivity test were performed. Because there was not more than 10 mmHg decrease in the mean pulmonary artery pressure (PAP) or 20% decrease in measured value of mean PAP, we considered the pulmonary reactivity test as negative.[7] Hence, staged surgical management was planned, and pulmonary banding was done, with the last correction being performed when the patient was three years old. To the best of our knowledge, the literature has described only five cases[1,3,8,9] of left hemitruncus and TOF that have survived beyond the first decade of life after successful surgical repair. This case is the first surviving patient to have undergone staged correction due to TOF with left hemitruncus while also having severe left pulmonary hypertension. As in our case, to prevent hemodynamically important pulmonary hypertension from occurring during cyanotic heart disease, pulmonary banding and pulmonary antihypertensive medication are commonly used.[10]

Early detection and repair of left hemitruncus is crucial because most children do not survive because of the early development of pulmonary vascular obstructive disease. When this condition is recognized in the early stages, surgical correction provides the best chance for survival.

Declaration of conflicting interests

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