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Case report / Olgu Sunumu

The simultaneous presence of bronchiectasis in a patient with double-outlet right ventricle, atrial septal defect, ventricular septal defect and pulmonary stenosis

Çift çıkımlı sağ ventrikül, atriyal septal defekt, ventriküler septal defekt ve pulmoner stenozu olan bir hastada bronşektazi birlikteliği

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Double-outlet right ventricle (DORV) is a very rare anomaly which can be presented with many cardiac abnormalities and ventricular septal defect (VSD) in particular. Clinical findings usually appear in the newborn period, which allows us to diagnose in the early period of life. The clinical findings vary according to the type of accompanying VSD and whether there are additional cardiac abnormalities such as pulmonary stenosis. These patients often present with cyanosis, congestive heart failure findings and pulmonary infections based on interstitial pulmonary edema. The treatment is surgical. Bronchiectasis is a serious pulmonary problem which may be encountered in some patients with frequent pulmonary infections. In this article, a 20-year-old male case with the simultaneous presence of bronchiectasis due to frequent pulmonary infections and DORV, VSD, pulmonary stenosis and atrial septal defect is presented. The risk factors of any pulmonary resection and cardiac surgery applied were also discussed.

Key words: Atrial septal defect; bronchiectasis; double-outlet right ventricle; pulmoner stenosis; ventricular septal defect.

Çift çıkımlı sağ ventrikül (ÇÇSV), başta ventriküler septal defekt (VSD) olmak üzere birçok kardiyak anomali ile birlikte görülebilen oldukça nadir bir anomalidir. Klinik bulguların genellikle yenidoğan döneminde gelişmesi nedeni ile yaşamın erken dönemlerinde tanı koyma olanağımız vardır. Klinik bulgular patolojiye eşlik eden VSD tipine ve pulmoner stenoz gibi ek kardiyak anomali olup olmamasına göre değişir. Bu hastalarda genellikle siyanoz, konjestif kalp yetmezliği bulguları ve interstisyel pulmoner ödem zemininde gelişen akciğer enfeksiyonları sık gözlenen klinik durumlardır. Tedavisi cerrahidir. Bronşektazi, sık akciğer enfeksiyonu gelişen hastalarda görülebilecek ciddi bir akciğer sorunudur. Bu yazıda, ÇÇSV, VSD, pulmoner stenoz ve atriyal septal defekt birlikteliği ve sık geçirilen akciğer enfeksiyonlarına bağlı olarak bronşektazi gelişmiş bir 20 yaşında erkek olgu sunuldu. Uygulanan kardiyak ameliyatlar ve uygulanabilecek herhangi bir akciğer rezeksiyonu sırasında ortaya çıkabilecek risk faktörleri de tartışıldı.

Anahtar sözcükler: Atriyal septal defekt; bronşektazi; çift çıkımlı sağ ventrikül; pulmoner stenoz; ventriküler septal defekt.

Double-outlet right ventricle (DORV) is a congenital cardiac anomaly characterized by the morphological exit of two big arteries from the right ventricle. It is seen equally between genders with an incidence rate of nine per 100,000 births. Furthermore, DORV may be seen together with many cardiovascular anomalies, but it is usually accompanied by a ventricular septal defect (VSD) since the only outlet for the left ventricle is through this.[1,2] Pulmonary edema and related frequent pulmonary infections may be seen in these patients, depending on the diameter and position of the accompanying VSD.[1,3] Bronchiectasis has many genetic, anatomic, and systemic causes, but the most common extrinsic developmental factor is frequent

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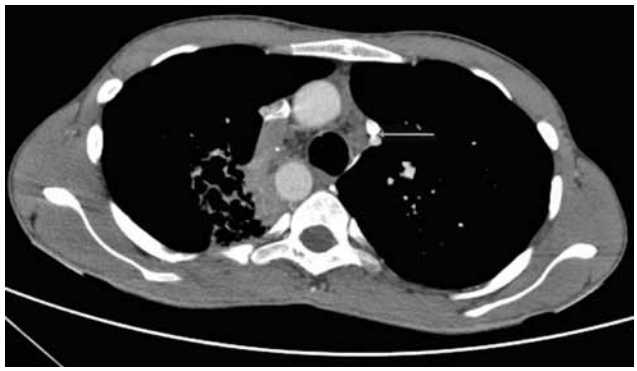

Figure 1. Right descending aorta (arrow).

pulmonary infections in childhood.[3] We present a case with bronchiectasis that showed concurrent DORV, VSD, pulmonary stenosis (PS), and atrial septal defect (ASD) along with a history of frequent pulmonary infections since childhood and then examine the various treatment options.

CASE REPORT

A 20-year-old male patient was found to have a murmur when he was 20 days old, and advanced investigations led to a diagnosis of simultaneous DORV, ASD, VSD, and PS. However, he had not been followed up. When he was seven years old, he was again admitted to the hospital with symptoms of fatigue and cyanosis. Angiography showed DORV, major artery transposition (Figure 1), PS, VSD, ASD, and right-sided arcus aorta. The Fontan operation was planned, but the patient's family refused any treatment. When the patient was 13, he presented at the hospital with symptoms of increasing cyanosis and fatigue. He underwent two systemic artery modified Blalock-Taussig (BT) shunt surgeries three years apart as a left BT shunt (Figure 2) was placed between the left subclavian and left pulmonary artery when the patient was 13 years old and a right BT shunt was inserted between the right subclavian and right pulmonary artery when he was 16. The patient began producing sputum when he was 13, and thoracic computed tomography (CT) showed widespread cystic bronchiectasis located centrally in the right upper lobe posterior segment, a hyperdense area in the right lung apex consistent with trapped air and focal bronchiectasis (Figure 3). In addition, there was the appearance of an adjacent budding tree in the right lower lobe superior segment and a shunt between the right pulmonary artery and vein. The patient had an oxygen (O_2) saturation rate of 88%, a partial pressure of oxygen (PO_2) rate of 56 mmHg, and a partial pressure of carbon dioxide (PCO_2) rate of 40 mmHg. Pulmonary function tests revealed a forced expiratory volume in one second (FEV1) of 2.24 (55% of the normal value) and a forced vital capacity (FVC) of 2.9 (63% of the normal value). A quantitative ventilation-perfusion scintigraphy of the patient showed that the right upper lobe functions had decreased to 3.7% as a result of the bronchiectasis, but there was no significant effect on the other areas (Table 1). A right upper lobectomy was planned accordingly.

Preoperative echocardiography showed a dilated coronary sinus, persistent left superior vena cava (SVC), vena azygos continuity, and a 2.5 cm wide VSD that could accept flow only through 1 cm. In addition there was the presence of DORV. Both atrioventricular (AV) valve orifices were present, but there was left AV valve regurgitation and atrial situs ambiguous. Pulmonary stenosis infundibular and valvular (max 60 gradient) BT shunt flow was present, and the major arteries were transposed (Figure 1). Atrial septal defect and an atrial septal aneurysm were also present. Preoperative angiography was planned, but the patient did not agree to it. Therefore, the angiography was not performed.

The right pulmonary venous return of the patient changed as a result of the performed surgery, and it

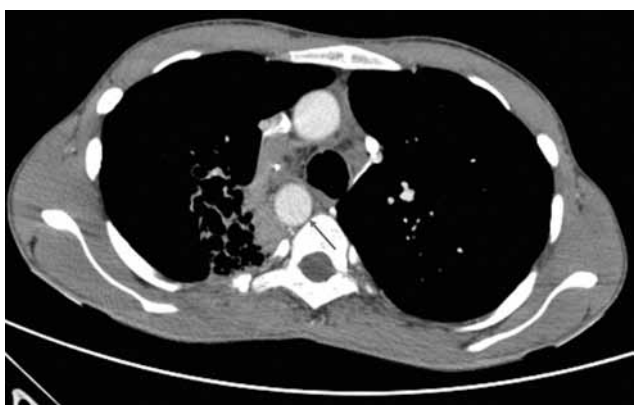

Figure 2. Left subclavian-left pulmonary artery Blalock-Taussig shunt (arrow).

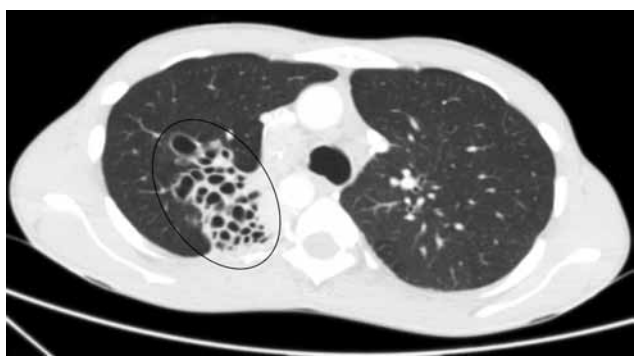

Figure 3. Focal bronchiectasis in the right lung apex.

Table 1. The differential functions of pulmonary scintigraphy

	Left	Right
Superior zone	13.8	3.7
Middle zone	28.8	23.1
Inferior zone	15.7	14.8

was believed that there would be a high risk of bleeding following a vascular ligation during any potential surgery. At that point, the patient was followed up and was also referred to a transplant center to be evaluated for a heart-lung transplant.

DISCUSSION

Double-outlet right ventricle is a rather rare congenital heart disease in which a ventriculoarterial connection anomaly occurs where both major arteries derive from the right ventricle morphologically. This anomaly can be accompanied by VSD, PS, major vessel abnormalities, and ASD as well as many other congenital heart diseases. Clinical findings depend on the type of accompanying VSD that is present and whether there is any additional cardiac anomaly, such as PS. Ventricular septal defect constitutes the most important part of the pathology in DORV since the only outlet of the left ventricle is through the VSD. Babies with this congenital disease usually develop congestive heart failure or cyanosis within the first two months of life on average and suffer from various clinical manifestations that need medical and surgical treatment, such as recurrent pulmonary infections due to pulmonary interstitial edema in later periods.[1,2]

Four types of VSD are found in patients with DORV, with the most common being the subaortic type. The clinical and hemodynamic findings are similar to those in the tetralogy of Fallot if pulmonary stenosis is present in DORV with subaortic VSD.[1,2] However, it is not always possible to differentiate between the two pathologies. Our case was diagnosed with subaortic VSD accompanied by ASD, PS, and DORV when he was 20 days old.

Bronchiectasis is a chronic inflammatory disease characterized by abnormal permanent dilatation of the bronchial wall,[3] and its underlying causes must be investigated when the diagnosis is made. Childhood pulmonary infections are the most important extrinsic cause of bronchiectasis. In fact, our patient had a history of frequent pulmonary infections with a productive cough that originated in his childhood. Concurrent DORV, VSD, PS, and ASD, as seen in our patient, causes a right-to-left shunt, and this increases the pulmonary blood flow and the systemic blood flow to higher than normal levels. The right ventricle becomes dilated, and the increased venous pressure in the lung causes interstitial edema, which leads to recurrent pulmonary infections. These infections, which start early in childhood, can cause bronchiectasis in some patients.[3] Bronchiectasis is most frequently seen in the left lower lobe (16.3%), but it can also involve the right upper lobe (6.3%),[3,4] as was the case in our patient. Acute attacks are treated with short-term systemic antibiotics, mucolytic agents, anti-inflammatory drugs, or bronchial hygiene therapy. The definitive treatment is surgery, and this should be the preferred option if the bronchiectasis is unilateral or shows segmental or lobar distribution, if it is progressive, or if the symptoms are recurrent.[5-7] The aim of surgical treatment is complete resection along with an increase in the patient's quality of life.[4,8] Surgical resection was considered in our patient because of the localized bronchiectasis and frequent recurrence of infection.

The concurrence of DORV and VSD is expected when we take into account the cardiac pathologies, and the added major artery transposition is actually an advanced form of DORV. A detailed investigation of these pathologies shows that PS protects the pulmonary arteries from pulmonary vascular disease. Our patient underwent a modified BT shunt two times within a three-year period between the ages of 13 and 16. His most recent echocardiography showed a McGoon ratio of over 1.5, so the case was deemed suitable for total correction.[2,7] However, the two working systemic artery-pulmonary artery modified BT shunts possibly would have most likely caused pulmonary hypertension again. A defining factor for this patient, for whom the Fontan operation had been planned, would have been the lack of pulmonary vascular disease development.[9,10]

However, pulmonary vascular disease had already developed as the maximum pulmonary valve gradient on echocardiography was 60 mmHg, and the patient was therefore considered to be high risk. A definitive diagnosis of pulmonary vascular disease can only be made with a pulmonary pressure measurement, but the patient refused angiography.

There are increased levels of pulmonary artery pressure with pulmonary artery clamping during surgery, and there are also more cases of cyanosis. The pulmonary pressure becomes even higher following a lobectomy and continues to have a noticeable effect on the cyanosis. Even if the pulmonary artery pressure is normal, there is a significant chance that it will increase postoperatively with the acceleration of the

pulmonary vascular disease development process, and this may make the Fontan operation impossible.[5,11] Another possible complication in our patient was the risk of blockage in the working BT shunts due to decreased flow following pulmonary artery clamping. A blockage would have led to decreased pulmonary flow and could have been fatal. The presence of a left innominate artery due to the right-sided arcus aorta pathology and the developed aortopulmonary collaterals due to PS would have made the surgery more complicated and could have caused excessive intraoperative bleeding. Therefore, we did not find it appropriate to operate on this bronchiectasis patient with complex cardiac anomalies. Hence, we put him on regular follow-up and gave priority a total correction along with a heart-lung transplant. Additionally, we provided medical and physical treatment as an inpatient during exacerbations.

In conclusion, a good preoperative evaluation is necessary for patients like ours who develop bronchiectasis on the basis of cardiac anomalies, and a conservative approach should be chosen if surgery is risky. Moreover, these patients should be placed under a heart-lung transplant evaluation program due to both their cardiac and pulmonary pathologies.

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