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Case Report / Olgu Sunumu

Rapid reversal of left ventricular enlargement after percutaneous closure of pulmonary sequestration in a newborn with prune belly syndrome

Prune belly sendromlu bir yenidoğanda pulmoner sekestrasyonun perkütan kapatılmasıyla sol ventriküler genişlemenin hızla düzelmesi

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ABSTRACT

In this article, we describe a newborn with Prune belly syndrome who presented with left ventricular dilation due to an extensive intralobar sequestration of the left lung. Pulmonary sequestration was combined with congenital cystic adenomatoid malformation and also had coarctation of the aorta. Percutaneous closure of the anomalous aberrant artery feeding the sequestrated lung and balloon angioplasty for coarctation resulted in prompt regression of the left ventricular enlargement in the catheterization lab.

Keywords: Aortic coarctation, bronchopulmonary sequestration, congenital cystic adenomatoid malformation, percutaneous intervention, Prune belly syndrome.

ÖZ

Bu yazıda sol akciğerde geniş intralober sekestrasyon nedeniyle sol ventrikül genişlemesi ile başvuran Prune belly sendromlu bir yenidoğan sunuldu. Pulmoner sekestrasyon, konjenital kistik adenomatoid malformasyon ile birlikteydi ve ayrıca aort koarktasyonu da eşlik ediyordu. Sekestre akciğeri besleyen anormal aberran arterin perkütan kapatılması ve koarktasyon için balon anjiyoplasti uygulanması, sol ventrikül genişlemesinde, kateterizasyon laboratuvarında hızla düzelme ile sonuçlandı.

Anahtar sözcükler: Aort koarktasyonu, bronkopulmoner sekestrasyon, konjenital kistik adenomatoid malformasyon, perkütan girişim, Prune belly sendromu.

The most common malformations of the lower respiratory tract are congenital cystic adenomatoid malformation (CCAM), known as congenital pulmonary airway malformation, and bronchopulmonary sequestration. Congenital cystic adenomatoid malformation of the lung is a rare congenital cystic lung lesion arising from excessive proliferation of tubular bronchial structures.[1] It is a disease of infancy, with most cases diagnosed within the first two years of life. It is a hamartomatous lesion containing tissue from different pulmonary origins. Bronchopulmonary sequestration is made of extraneous and non-functioning lung tissue separated itself from the standard pulmonary structure.[2]

Prune belly syndrome is composed of a characteristic clinical triad: abdominal wall muscle deficiency, undescended testes, and urinary tract malformations. Other anomalies such as CCAM are also associated with this syndrome. The prognosis is poor and many patients die at an early age of urinary tract infection, renal failure, or pulmonary complications.[3]

In this article, we report a newborn with CCAM whose clinical condition improved, pulmonary arterial pressure decreased immediately after transcatheter embolization of anomalous systemic arterial supply and balloon angioplasty for coarctation of the aorta.

CASE REPORT

A full-term male patient was born with a birth weight of 2.8 kg. He was admitted to the neonatal intensive care unit due to respiratory distress and intubated. Computed tomography (CT) was performed in the patient who could not be weaned from the mechanical ventilator. The CT showed aortic coarctation, patent ductus arteriosus (PDA), and CCAM at the left lung

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and two sequestration arteries, feeding the left lung's inferior lobe (Figure 1). Based on these findings, he was referred to our hospital on Day 12. On our patient's physical examination, the abdominal wall was bulging out in the flanks area, and he had bilateral cryptorchidism. The abdominal ultrasonography showed mild dilatation of the ureters characterizing hydroureteronephrosis and the kidney function was normal. By transthoracic echocardiography (TTE), dextroversion was revealed due to the mediastinal shift. The pulse-Doppler trace of the descending aorta demonstrated a continuous flow during the cardiac cycle with a 40-mmHg peak gradient supporting the aortic coarctation. Enlarged left cardiac chambers, moderate mitral regurgitation, PDA with the bidirectional flow were also detected.

Therefore, we decided to perform a cardiac catheterization to close the sequestration arteries percutaneously and interfere with aortic coarctation. Angiography confirmed the presence of two large anomalous arteries, having diameters of 3.5 mm and 3 mm, which arose from the descending thoracic aorta and fed two sequestrated inferior lobes of the left lung (Figure 2a). The venous return phase showed the veins of these two lobes returned to the left atrium. A 4-Fr Cook long sheath (Cook, Bloomington, IN, USA) was placed into the sequestration artery. Then, a 6-mm Amplatzer Vascular Plug-4 (AVP-4) (AGA Medical, MN, USA) was implanted first, and an 8-mm AVP-4 was implanted into the other sequestration artery. Following injections showed complete occlusion of the sequestration (Figure 2b). Pulmonary artery pressures

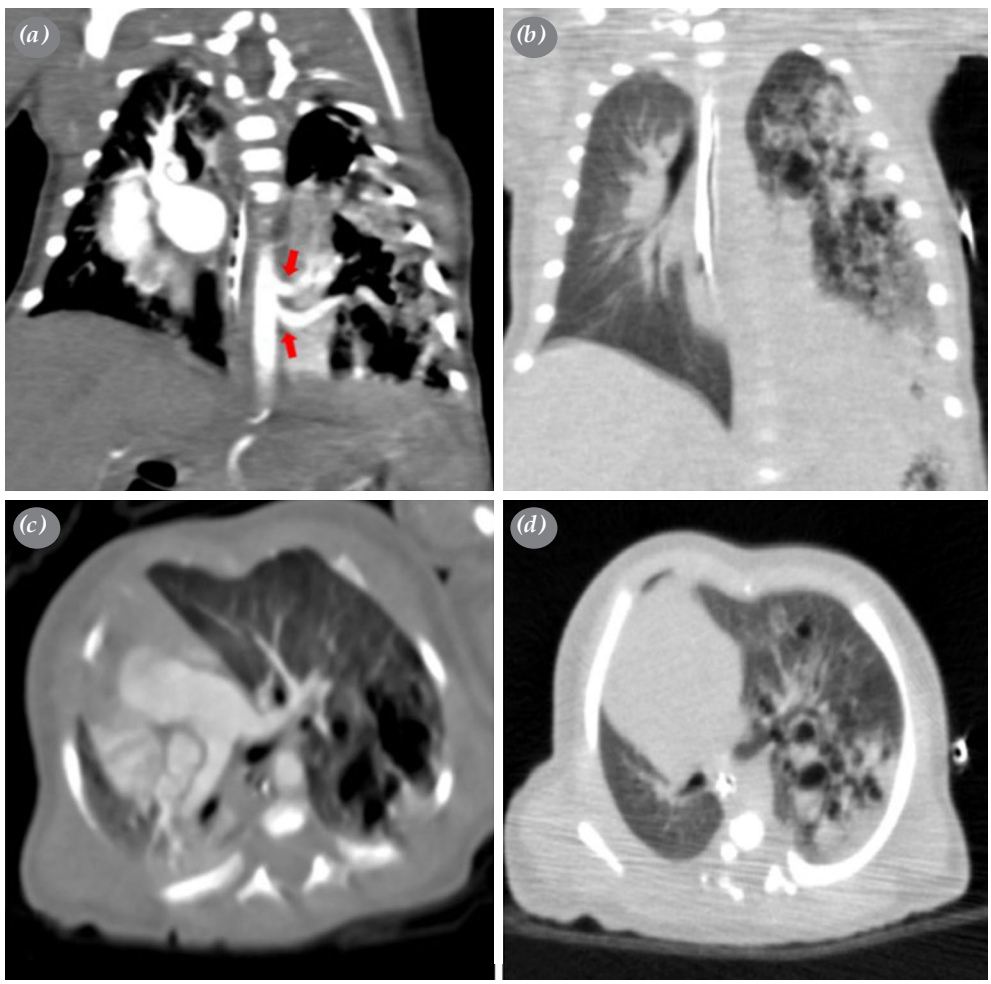

Figure 1. Computed tomography angiography demonstrating. **(a)** The vascular anatomy of the two feeding arteries (red arrows), **(b)** multiple small cystic lesions in the left lung at coronal section, **(c)** Pulmonary arteries with cystic lung lesion in the left lobe with two systemic arteries originating from the thoracic aorta, **(d)** Small cystic lesions with consolidation in the left lung at axial section.

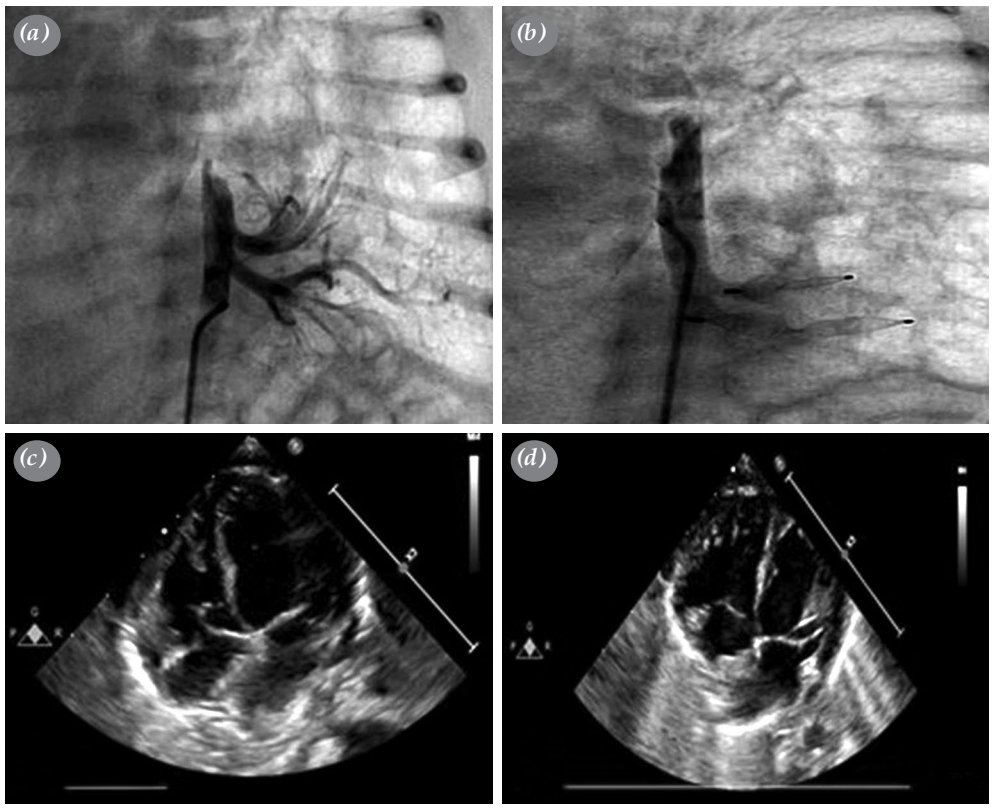

Figure 2. (a) Conventional angiography showed two sequestration artery originated from descending artery. **(b)** Complete occlusion of the sequestration after vascular plugs. **(c)** Transthoracic echocardiography in the apical 4-chamber view demonstrating dilation of left heart chambers before the intervention. **(d)** Regression of left ventricular dilation immediately after intervention.

were measured as 46/33 (39) mmHg during the procedure.

In the arcus injections, the distal aortic arch was 3.3 mm and coarctate segment was 2.2 mm in the isthmic region, and the descending aorta was 4.6 mm. Then balloon angioplasty was applied with a 4×20 mm Tyshak® balloon to the coarcted segment over nitinol wire. Before the procedure, the ascending aorta pressure was 69/27 (45) mmHg, while the descending aorta was 47/28 (38) mmHg, with a 22-mmHg gradient. After the procedure, the ascending aorta pressure was 44/17 (28) mmHg, and the descending aorta was 43/20 (30) mmHg, and the peak-to-peak gradient was almost disappeared.

The TTE performed 30 min after the procedure showed that mitral regurgitation was healed to a minor degree, the enlargement in the left spaces returned to normal, and the PDA shunt turned from left to right (Figure 2c, d). There were no complications after catheterization, and the patient was discharged uneventfully without requiring supplemental oxygen. The patient was followed in the intensive care unit for one day and, then, in the inpatient pediatric unit for two days. He was transferred to the clinic, where he was referred to our center and was discharged from there three days later.

DISCUSSION

Congenital bronchopulmonary malformations are rare. Bronchopulmonary sequestration and CCAM are well-known entities. Approximately 60% of CCAM cases are associated with other findings such as cardiac anomalies, renal agenesis/dysgenesis, gastrointestinal atresia, and skeletal anomalies. In the CCAM, the pathological changes occur approximately at the fifth or sixth week of gestation. Clinically 85% of infants with CCAM present in the neonatal period with respiratory problems and, it is slightly more common in males. If symptoms are subtle in the newborn period, progressive respiratory distress and infection may occur later in infancy. Bronchopulmonary sequestrations are microscopic cystic masses of non-functioning

pulmonary tissue that lack an obvious communication with the tracheobronchial tree. In bronchopulmonary sequestration, the lung tissue receives all or most of its blood supply from an anomalous systemic artery, with this artery's origin being variable.[1] Our patient was male, had a unilateral type 2 CCAM and intralobar bronchopulmonary sequestration, associated with aortic coarctation as a cardiac anomaly and PBS. Congenital bronchopulmonary malformations and PBS are unfortunate combinations. Compression of adjacent lobes, displacement of the heart, and mediastinum toward the opposite side of the chest cause respiratory difficulties in these patient groups.[3] The sequestered lobe may cause substantial arteriovenous shunting, leading to high-output cardiac failure, as in our patient. Surgical or percutaneous methods are recognized for the treatment in the neonatal period. Wu et al.[4] reported an extensive series of 340 patients diagnosed with CCAMs. They recommend conservative management for all children aged <6 months with CCAMs, if no symptoms are present. If the child develops more than two bouts of pneumonia within six months or symptoms appear after six months, an operation is advised. An increased risk of infection in children of kindergarten age or malignant transformation after three years of age should be concerned. Due to the high incidence of complications such as pneumothorax and mediastinal shift, type 4 CCAMs, which is characterized by peripheral cysts that originate in alveolar cells, may be resected earlier than other cystic CCAMs. The standard of care from the lung lesions is a lobectomy, but segmentectomy and embolization alone have been reported.[4,5] In percutaneous methods, to the best of our knowledge, the Amplatzer™ Vascular Plugs are known as effective and safe devices with different size choices.

In conclusion, since the coexistence of CCAM and bronchopulmonary sequestration is a rare occurrence, we report a male infant who had type 2 CCAM combined with intralober bronchopulmonary sequestration. Their combination with the coarctation of the aorta may easily improve symptoms of heart failure and pulmonary hypertension in infants. Percutaneous closure of the sequestration arteries and treating associated anomalies produce a simultaneous reversal of the left ventricular enlargement.

Patient Consent for Publication: A written informed consent was obtained from the parents of the patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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