Right pulmonary artery – left atrial communication presenting with brain abscess: a case report

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SUMMARY: Küçükosmanoğlu O, Poyrazoğlu H, Topçuoğlu MŞ, Erdem S, Erman T, Özbarlas N. Right pulmonary artery-left atrial communication presenting with brain abscess: a case report. Turk J Pediatr 2006; 48: 373-375.

Direct communication between the right pulmonary artery and left atrium is a very rare vascular malformation. We report a patient with this anomaly. She presented with unexplained cyanosis and brain abscesses. The diagnosis was made with contrast echocardiography and angiography. We treated this anomaly successfully with surgery. Complete cure for this anomaly can be achieved by ligation.

Key words: cyanosis, right pulmonary artery-left atrial communication, cerebral abscess.

Direct communication between the right pulmonary artery (RPA) and left atrium (LA) is a very rare congenital malformation[1,2]. The first case was reported by Friedlich et al. [3] in 1950. Since then 54 cases have been reported in the English literature. The amount of right-to-left shunt via RPA-LA communication establishes the severity of the symptoms and age of clinical presentation. Clinical presentation of a large communication consists of central cyanosis with decreased oxygen saturation, tachypnea and respiratory distress in the neonate. Older patients had milder symptoms and significant delay in diagnosis. The age at diagnosis ranged from 1 month to 60 years. Cerebral abscess was noted as a possible complication in adolescents and adults. Herein, we report a case of RPA-LA communication presented with brain abscess.

Case Report

A 16-year-old cyanotic girl was referred to our neurosurgery department because of brain abscess. According to her history, she had been cyanotic since her early infancy but had not been evaluated as a cardiac patient. Physical examination revealed marked central cyanosis and clubbing. Heart sounds were normal. A 12-lead ECG showed mild left atrial hypertrophy. A 2-D echocardiography and color Doppler studies were normal. In a contrast echocardiography, early filling of the LA with contrast material was seen. This suggested presence of a pulmonary arteriovenous fistula and cardiac catheterization was planned. Computed tomography showed two abscesses in the right temporal and left frontal lobe. Brain abscesses were resolved with surgical drainage and antibiotics. In postoperative 14th day, she underwent cardiac catheterization. The cardiac catheterization revealed normal right atrial, right ventricular, pulmonary artery, and left atrial, left ventricular and aortic pressures.

Oxygen saturations were as follows: 49-51% in the right atrium, the right ventricle and the pulmonary artery, 77-79% in the LA, left ventricle, and the aorta, and 97% in the pulmonary vein. The Qp/Qs was calculated as 0.62:1. Selective left pulmonary artery angiogram revealed a normal left pulmonary artery tree and normal left side pulmonary venous drainage. RPA angiogram revealed a large tunnel-like vessel approximately equal in size to proximal RPA, which arose from the posterior side of the RPA and entered the right side of LA (Fig. 1). The distal RPA tree and pulmonary venous drainage were normal.

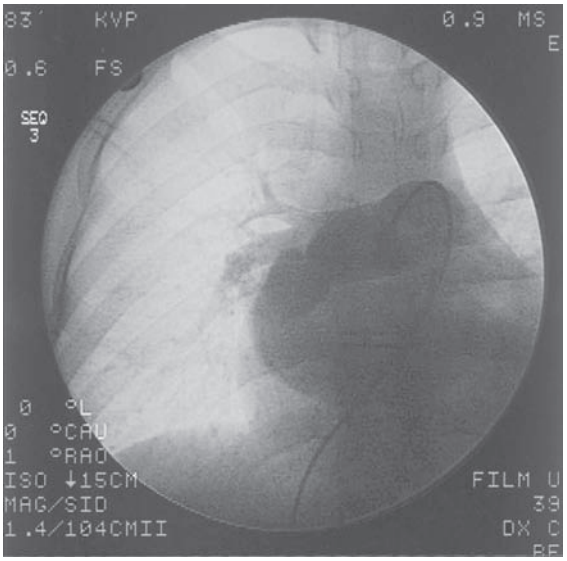

Fig. 1. Posteroanterior view of selective angiocardiogram. Right pulmonary artery (RPA) revealed a large tunnel-like vessel which arose from the posterior side of RPA and entered the right side of the left atrium (LA).

When the NIH catheter was pushed into the tunnel, it was easily passed to the LA and then to the left ventricle. We also performed the left ventricle and LA angiograms while the catheter was located in the tunnel (Fig. 2).

After four weeks of intravenous antibiotic therapy, the patient underwent an operation for her cardiac anomaly. Surgery was performed through a median sternotomy without cardiopulmonary by-pass. Upon dissecting the RPA, an anomalous vascular structure arising from its posterior was seen (Fig. 3). The vessel was traced and seen to enter the LA posteriorly. On clamping the communication, the oxygen saturation rose to 98%. The tunnel was closed with simple ligation and transfixion techniques. The patient became acyanotic with arterial oxygen saturations of 98% after surgery. The girl's postoperative course was uneventful and she remained asymptomatic during the 20 months of follow-up since the operation.

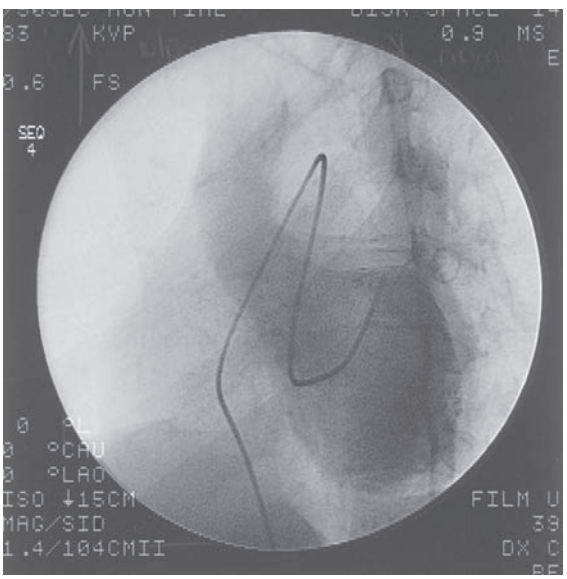

Fig. 2. The left ventricle angiogram was performed while the catheter was located in the tunnel.

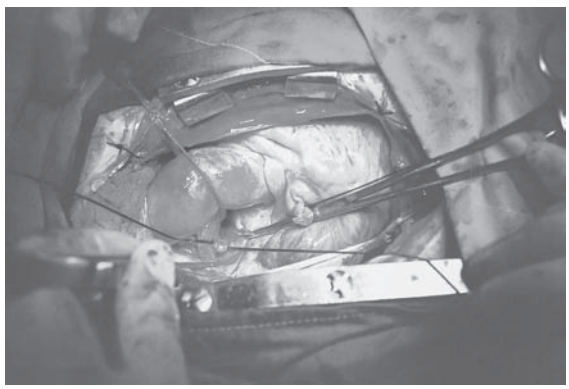

Fig. 3. Intraoperative view of the right pulmonary artery (RPA) demonstrating an anomalous vascular structure arising from its posterior.

Discussion

According to de Souza e Silva et al.[4], communication between the RPA and the LA can be classified into three types based upon the anatomy of pulmonary venous drainage pattern. An additional fourth type was later described by Ohara et al.[5]:

Type I: Normal pulmonary venous drainage pattern.

Type II: Absent right inferior pulmonary vein with fistulous connection at the normal site of its origin.

Type III: All pulmonary veins connected to the aneurysmal pouch.

Type IV: Right inferior pulmonary vein replaced by three small veins connected to the aneurysmal pouch.

Our patient had Type I communication. Because of the low socioeconomic status of our patient, the diagnosis was delayed. Absence

of symptoms of cardiac failure also had an important role in the delayed diagnosis. Surgical ligation is the accepted treatment for RPA-LA communication, but transcatheter coil occlusion for small communication[6] and Amplatzer device occlusion for larger ones have been described as alternative treatments[7].

Although it is rare, RPA-LA communication is an important cause of central cyanosis and the complications of cyanotic cardiac disease such as brain abscess. If transthoracic echocardiography is normal, differential diagnosis of this anomaly should be made in the cyanotic patient, especially with brain abscess. The clinical diagnosis of this anomaly is difficult. The transthoracic echocardiography may be normal. Contrast echocardiography is a simple and easy diagnostic investigation for diagnosis of pulmonary arteriovenous fistula. Positive contrast echocardiography suggested pulmonary arteriovenous fistula, but not the level of the fistula. Cardiac catheterization with selective angiography of the RPA is necessary to confirm this unusual diagnosis.

In conclusion, if transthoracic echocardiography is normal in the patient with central cyanosis, contrast echocardiography should be made for differential diagnosis of this vascular anomaly.

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