Introduction

Isolated pulmonary artery agenesis is observed at a rate of 1/200,000 (1). The great majority of the cases in the literature have been diagnosed in infancy and childhood. Shortness of breath, coughing, and frequent pulmonary infections have been the symptoms leading to diagnosis. Diagnoses at later ages have been made coincidentally after suspecting the chest X-rays taken.

Case Report

Our case was a 22-year-old male patient admitted to our out-patient clinic with the complaint of shortness of breath upon exertion that started 15 days ago. His personal history included smoking 16 packets / year while his family history did not have anything worth noting.

In his physical examination, he was observed to be in good general condition, fully conscious and co-operative, and his vital signs were natural. Upon examination of the respiratory system, the patient’s left hemithorax’s participation in respiration was limited and upon auscultation decreased lung sounds were present. Cardiac sounds were rhythmic and there were no additional sounds.

In the PA lung x-ray of the patient, the volume of the left hemithorax was observed to be decreased; the mediastinum was deviated to the left and the left lung has hypoplasia appearance (Fig. 1).

Kaynaklar

In the computed tomography of the thorax the left pulmonary artery was observed to be absent (Fig. 2). Since the patient did not accept to undergo a bronchoscopic procedure, a virtual bronchoscopy was performed to understand whether there was an endobronchial lesion and to elucidate the bronchial anatomy. All bronchial and segment openings were observed to be open and there were no endobronchial lesions (Fig. 3).

No pathology was detected in the respiratory function test. In order to make a definite diagnosis, a pulmonary digital subtraction angiography was performed on the patient by entering from the right femoral artery and left pulmonary artery agenesis was observed in the angiography (Fig. 4).

Since agenesis of the left pulmonary artery is known to co-exist very frequently with cardiac anomalies, echocardiography was performed to exclude cardiac pathology, and no cardiac and vascular anomalies were observed. The patient’s pulmonary artery pressure at rest was measured as 20 mm Hg.

Discussion

Congenital pulmonary artery agenesis is a rare anomaly that frequently co-exists with cardiac anomalies (1).

Embryologic data; Before the development of the pulmonary artery, the lung is supplied by small transitory branches of the dorsal aorta that later disappear. These branches persist as enlarged supradiaphragmatic and infradiaphragmatic aortic branches, or as bronchial arteries in cases in which the pulmonary artery does not develop (2).

The first case in the literature was detected in 1868 (3) and two more reviews were published in 1962 and 1978 (4, 5). With the most recent review published in 2002 a total of hundred and eight cases of isolated pulmonary artery agenesis accompanied by cardiac anomalies have been reported (1).

Fourteen of these cases were asymptomatic and the average age of detection was 14. The patient in our case was 22 years old and compared to other cases in the literature was at a relatively older age. In 37 percent of the 108 cases there was a history of frequent pulmonary infections.

In our case, there was no history of frequent pulmonary infections. While pulmonary hypertension was detected in 25% of the cases, the pulmonary artery pressure in our case was 20 mm Hg. While right pulmonary artery agenesis was detected in 63% of the cases in the last review, in our case the rarely encountered left pulmonary artery agenesis was detected. Pulmonary edema was observed in 12% of these 108 cases and in some of them respiratory insufficiency developed in the later stages. In the high-resolution computerized tomographies of the thorax taken in some cases because of a history of frequent pulmonary infections minimal or widespread bronchiectasis was diagnosed (1). In the thorax tomography taken in our case, on the other hand, there was no bronchiectasis. Exercise limitations were described in 40% of the 108 cases and it was also determined in our case with the same symptomatology.

Most of the cases in the literature were diagnosed with the development of recurrent pneumonia. Some of the cases in the literature were detected coincidentally during screening for tuberculosis. There is
also a publication indicating that a good tomography interpretation is often adequate in detecting pulmonary artery agenesis (6).

In a literature publication from 1982; a series comprised of 19 cases was examined and ligation was performed on two patients because of frequent hemoptysis attacks (7). In a case report in 1993; pulmonary hypertension was detected in two cases from a series comprised of 4 cases and both of them died in two years. The most important complications affecting the survivals of these cases were; pulmonary hypertension and frequent infections.

In a literature publication from 2004, the average age of the eight cases detected was 11, and four of them were followed up until adult years and no complications were seen (8). One case of pulmonary artery agenesis with stenosis of the main bronchus that was diagnosed after being admitted to the out-patient clinic with asthma-like attacks was also reported (9). A patient with pulmonary artery agenesis who has shortness of breath and pleuritic chest pain was admitted with the classical symptomatology of the condition and diagnosed (10).

Our case had not shown any symptoms previously either and was diagnosed with this condition after the age of 20 upon shortness of breath with effort. Since the survival rates of patients diagnosed at younger ages are increased due to operations for complications, we may suggest that our case’s not being symptomatic until his adult age will not affect his survival rate.

Conclusion
We wanted to present our case because it was an isolated case, the condition could be observed less frequently in the left pulmonary artery, it was detected at a rather late age, it did not have pulmonary hypertension, it did not follow a course with complications and it did not have any additional cardiac anomaly.

References

Fibroelastoma of the posterior mitral leaflet

Posteriyor mitral yaprakçıkta fibroelastoma

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Introduction
A papillary fibroelastoma is a very uncommon primary tumor of the heart with benign pathology. The tumor is generally diagnosed accidentally during echocardiography or following the investigation of a thromboembolic patient (1, 2). Valvular involvement includes aortic and mitral valves. Most of the scarce data have been reported from patients older than 50 years old, it may be found at any age, from the neonate to the nonagenarian (2). We present a rare case of papillary fibroelastoma of the posterior mitral leaflet in a 52-year-old patient.

Case report
A 52-year-old lady was referred to our clinic with palpitation and dyspnea and a mobile mass was demonstrated with transthoracic echocardiography on the posterior leaflet of the mitral valve. She had no prior history of a thromboembolic event. The transeosophageal echocardiography (TEE) demonstrated a sessile tumor 10 mm in diameter and mild mitral regurgitation (Fig 1A. and 1B). It is noteworthy that the mitral annular diameter was 26 mm on echocardiography. The surgical removal of the tumor was planned. Following standard