

# Multislice CT Findings of Left Pulmonary Artery Agenesis in an Adult Patient: Case Report

## Sol Pulmoner Arter Agenezisli Erişkin Hastada Çok Kesitli BT Bulguları

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**ABSTRACT** The congenital absence of pulmonary artery is a rare result of embryologic malformation and it could be associated with other anomalies such as cardiovascular and/or skeletal anomalies. A patient with left pulmonary artery agenesis with major aortopulmonary collateral arteries was discussed by the findings detected at multislice computed tomography. To the best of our knowledge, our patient is one of the oldest cases that were diagnosed as pulmonary artery agenesis, which is an extremely rare condition in adults. Recurrent pulmonary infection, hemorrhage, and mild dyspnea in exercise are the most common symptoms. One third of patients are asymptomatic, while approximately 50% of them would experience recurrent chest infections. In our patient left pulmonary artery agenesis was combined with right aortic arcus, and with the collaterals from inferior phrenic artery, thoracic arteries, left internal mammary artery and thyrocervical artery.

**Key Words:** Pulmonary artery; abnormalities; tomography, spiral computed

**ÖZET** Pulmoner arterin yokluğu nadir bir embriyolojik malformasyon sonucu olup bu duruma kardiyovasküler ve/veya iskelet sistemi anomalileri gibi diğer anomaliler eşlik edebilir. Bu sunumda sol pulmoner arter agenezisli hasta ve bu hastanın major aortopulmoner kollateral arterleri çok kesitli BT bulguları ile birlikte tartışılmaktadır. Bilgilerimize göre sunduğumuz olgu, literatürdeki en yaşlı olgulardan biri olup erişkinlerde saptanması son derece nadirdir. Rekürren pulmoner enfeksiyon, kanama ve hafif egzersiz dispnesi en sık semptomlardır. Olguların üçte birinde asemptomatik iken hastaların neredeyse yarıya yakınında rekürren pulmoner enfeksiyon gelişir. Sunduğumuz hastada sol pulmoner arter agenezisi ile birlikte sağ aortik ark ve inferior frenik arterden, torasik arterlerden, sol internal mamaryan arterden ve tiroservikal arterden gelen kollateral vasküler yapılar da mevcuttu.

**Anahtar Kelimeler:** Pulmoner arter; anormallikler; tomografi, spiral bilgisayarlı

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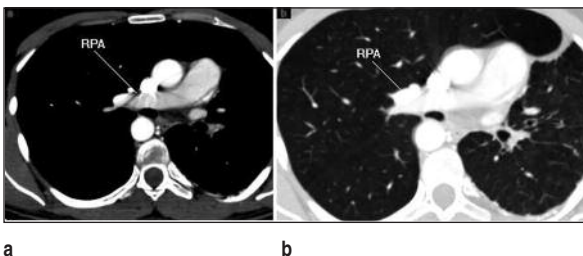
The congenital absence of pulmonary artery is a rare result of embryological malformation and it could be associated with other anomalies such as cardiovascular and/or skeletal anomalies. The incidence of unilateral pulmonary artery agenesis (UPAA) is 1/200 000. The great majority of the cases in the literature have been reported to be diagnosed in infancy and childhood. The unilateral absence of right pulmonary artery is more common than the left one; in the other hand UPAA is frequently more complicated and associated with cardiovascular anomalies.<sup>1,2</sup>

We introduced a patient with left pulmonary artery agenesis, in who collateral circulations called major aortopulmonary collateral arteries

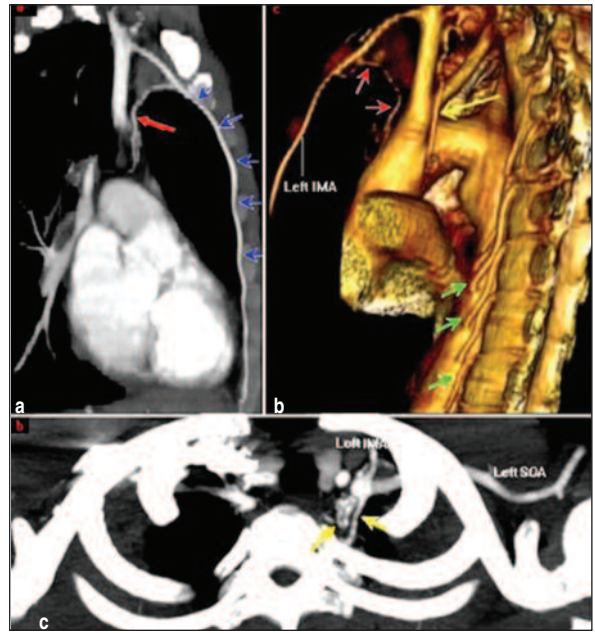
(MAPCA) exist. The diagnosis was supported by the findings detected at Multi Detector Computed Tomography (MDCT).

## CASE REPORT

A 32-year-old male patient was admitted to our hospital with the complaint of chest pain and dyspnea. There was no significant finding in his physical examination and laboratory results did not reveal a significant abnormality. Electrocardiogram (ECG) and functional respiratory tests were normal. In the posterior-anterior (P-A) chest roentgenogram of the patient, the volume of the left hemitorax was decreased, the heart, the mediastinum and the trachea were deviated to the left side, and the arcus aorta was not visible (not shown). With these findings a vascular abnormality was thought and MDCT angiography were planned. In MDCT images (Toshiba Aquillion 64 detector, Tokyo, Japan), the volume of the left lung was significantly decreased when compared to the right lung, apex of right lung was herniated to the left side and air was trapped in the herniated part of the right lung (Figure 1a). Pulmonary truncus and right pulmonary artery were calibrated normally. Arcus aorta was located at the right side. In addition, the left pulmonary artery could not be seen from its level of origin (Figure 1b). We observed four collaterals (MAPCAs) in this patient. The first MAPCA was from the left internal mammary artery (IMA) (Figure 2a, c), second and third ones were from thyrocervical branch of the left subclavian artery (SCA) (Figure 2b, c), and fourth MAPCA was a col-



**FIGURE 1:** **1a.** In axial CT scan in mediasten window settings; the left lung volume was significantly decreased and apex of right lung was herniated to the left side. Heart and mediastinum were also deviated leftward. **1b.** Axial CT scan in lung window settings of the same level also showed absence of the left pulmonary artery. Right pulmonary artery (RPA) diameters were in normal ranges.



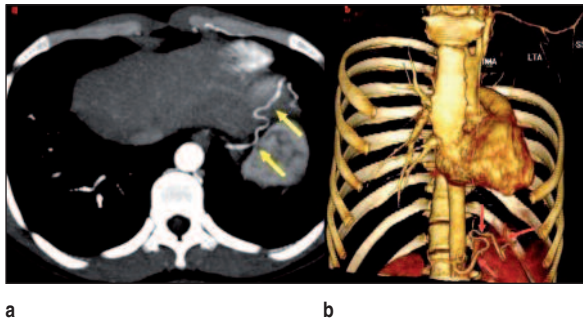
**FIGURE 2:** **2a.** Sagittal MIP image shows MAPCA (red arrow) from dilated left IMA (blue arrows). **2b.** Axial MIP image shows MAPCAs (yellow arrows) originating from the thyrocervical branch of the left SCA. **2c.** Sagittal VR image shows MAPCA from left IMA (red arrows), MAPCA from thyrocervical branch (yellow arrows) and dilated intercostals arteries (green arrows).

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lateral from left inferior phrenic artery (IPA) (Figure 3a, b). The first MAPCA from the left IMA supplied superior segmentary pulmonary artery branches and the fourth MAPCA from the left IPA supplied inferior segmentary pulmonary artery branches. Second and third MAPCA's from thyrocervical branch of the left SCA supplied both superior and inferior pulmonary artery branches entering medial side of the lung. MDCT clearly showed MAPCAs in this patient. In addition, we noted dilated intercostal arteries arising from the lateral side of the descending aorta (Figure 2c). The heart size was in normal ranges. Because of the risk for association of the cardiovascular abnormalities transesophageal echocardiography was performed; there was no abnormality observed in heart and big vessels but right arcus aorta.

## DISCUSSION

UPAA is a rare condition which is a result of malformation of the 6<sup>th</sup> aortic arch at the affected side. It could be an isolated condition but it is usually associated with cardiovascular abnormalities such as



**FIGURE 3:** Axial MIP image (a) and coronal VR image (b) show the collateral originating from the left inferior phrenic artery (arrows). Arcus aorta is right sided. VR image also shows left internal mammary (IMA), lateral thoracic artery (LTA) and SS (subscapular artery).

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tetralogy of Fallot, right arcus aorta, septal defect, patent ductus arteriosus, coarctation of aorta, subvalvular aortic stenosis, and big vessel transposition.<sup>3</sup> In patients with UPA, the intrapulmonary vessels and distal segment of the affected pulmonary arterial truncus normally develop and take the blood supply from the collaterals such as bronchial, intercostal, subclavian and subdiaphragmatic arteries. These collaterals enter into the lung and supply the segmenter pulmonary arteries. Only in 3 cases the collaterals were reported to be originated from coronary arteries.<sup>2</sup>

The first pulmonary artery agenesis case in the literature was presented in 1968 and 108 cases have been reported until year 2002. Mean age of those cases was reported to be 14 years. Forty percent of them were reported to have recurrent pulmonary infection and 25% of them pulmonary hypertension, and both of these complications affected the mortality rate strongly.<sup>3</sup>

The patients with right pulmonary artery agenesis might be asymptomatic for a long period. Recurrent pulmonary infection, hemorrhage, and mild dyspnea in exercise are the most common symptoms. One third of patients are asymptomatic, while approximately 50% of them would experience recurrent chest infections. Our patient had no history of recurrent pulmonary infection. These recurrent infections might be related to hypoperfusion. Shortness of breath during exertion, and reduction of exercise toleration are common find-

ings.<sup>2</sup> Pulmonary hypertension develops in only a small proportion of the patients.<sup>4</sup> Since these symptoms are not characteristic; the diagnosis of this condition could be delayed or mistaken.

Chest roentgenograms show a volume loss in the affected hemithorax, nevertheless diaphragmatic elevation with shifting of the heart and mediastinum to the affected side is detected most commonly and the contralateral lung is hyperinflated and herniated into the affected hemithorax as in our case. If there are enlarged intercostal and transpleural arteries, fine linear opacities could be seen at the periphery of the lung. Though our case have enlarged intercostals arteries, no parenchymal changes were observed.

On CT scans, the mediastinal portion of the affected pulmonary artery is often completely absent or terminates within 1 cm of its origin.<sup>4,5</sup> In our case left pulmonary artery was completely absent.

Serrated thickening of the pleura and subpleural parenchymal bands might be observed on CT scans and could be attributable to direct anastomosis of transpleural collateral vessels with peripheral branches of the pulmonary artery. Agenesis or hypoplasia of pulmonary artery could be detected easily with CT and MR.<sup>5</sup>

Swyer-James Macleod's (SJM) syndrome might be considered for differential diagnosis. SJM syndrome is a subtype of bronchiolitis obliterans, it might occur as a result of pneumonia in childhood. Radiologic findings of SJM syndrome are, hyperinflated and normal/decreased sizes of the affected lung and reduction of the vascularity and air trapping in expiration, with no obliteration in main branches.<sup>6</sup> There is no absence of any pulmonary artery in these cases in contrast to our case.

In case of abnormally originated pulmonary artery, one or both pulmonary arteries may arise from ascending aorta or directly from aortic arcus. In 80% of these patients the abnormal originated artery is the right one and 75% of them have patent ductus arteriosus.<sup>7</sup>

Another disease that must be considered in the differential diagnosis is syndrome of Scimitar,

which can be described as the abnormal development of the lung. The right side is affected more frequently than the left side. The affected lung is small and hyperlucent due to hypoplasia or absence of the pulmonary artery. Systemic collaterals supply blood to the affected lung. It could be diagnosed by observing the abnormal vein that drains the affected lung. This abnormal vein is named as Scimitar vein and it drains to the vena cava inferior.<sup>8</sup>

MDCT technology improved the capability of diagnosis in vascular pathologies as in our case. A large field of volume (FOV) can be scanned in a few seconds. 32 cm (with 0.5 mm slice thickness) can be covered with one gantry rotation time with 64 detector CT. Low scan time and increased z-axis resolution (isotropic imaging) with MDCT also made 3-D tools, MPR (multiplanar reformat), MIP (minimum and maximum intensity projection) and VR (volume rendering) possible which are very important in the diagnosis.<sup>9</sup> Dilated collateral arteries, their origins and anastomoses with other systemic arteries are easily detected without the

need for DSA (digital subtraction angiography) which is an invasive procedure. In our case we observed 4 MAPCAs. One from dilated left IMA, two from thyrocervical branch of the left subclavian artery (SCA) and one from the left inferior phrenic artery. MDCT also demonstrated dilated left intercostals arteries.

Magnetic Resonance (MR) has lost the advantage of having the only radiologic modality that is capable of multiplanar imaging. But MR still has some other advantages that, it uses no X-Ray, etc.

## CONCLUSION

To the best of our knowledge, our patient is one of the oldest cases that were diagnosed as pulmonary artery agenesis, which is an extremely rare condition in adults. In our patient left pulmonary artery agenesis was combined with right aortic arcus, and with the collaterals from inferior phrenic artery, left IMA and thyrocervical artery called MAPCA. MDCT angiography can demonstrate this abnormality and associated conditions thoroughly.

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