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Anomalous Orıgın of the Left Pulmonary Artery from the Ascendıng Aorta wıth Pulmonary Atresıa ın a 13 Year-old Girl

Tuba Selcuk[1,2*], Cigdem Bilgili[1], Hafize Otcu[3], Yildiray Savas[1], Gulfidan Cakmak[4] and Ferhat Cengel[1]

[1]Department of Radiology, Istanbul Haseki Education and Research Hospital, Haseki, Istanbul, Turkey.
[2]Department of Medical Sciences, Gelisim University, Avcılar, Istanbul, Turkey.
[3]Department of Radiology, Reyhanlı Government Hospital, Reyhanlı, Hatay, Turkey.
[4]Department of Pulmonary Medicine, Istanbul Haseki Education and Research Hospital, Haseki, Istanbul, Turkey.

Authors' contributions

This work was carried out in collaboration between all authors. Author TS designed the study, wrote the protocol and the first draft of the manuscript. Author CB performed the statistical analysis. Authors HO and YS managed the analyses of the study. Authors GC and FC managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aim: We reported a rare case of an anomalous origin of the left pulmonary artery (AOLPA) from the ascending aorta associated with pulmonary atresia and right sided aortic arch diagnosed at a relatively late age.
Case: 13 year-old girl presented to our pulmonology clinic with complaints of cough and dyspnea. On chest X-ray cardio thoracic ratio was increased and shadow of the arch was not seen on the left. On her echocardiography pulmonary arteries couldn't be demonstrated. Computed tomography

**Corresponding author: Email: drtubas@gmail.com;*

angiography was performed to the patient. Right sided arch aorta with pulmonary atresia associated with an anomalous origin of the left pulmonary artery from the ascending aorta with a well developed collateral blood supply to the right lung and coexisting pulmonary infection was detected. She was managed medically. She is on the first year of her follow up. Her medical status is stable.
Conclusion: We presented a case of relatively rarely seen anomalous origin of the left pulmonary artery from the ascending aorta with a rarely seen association of pulmonary atresia and wanted to take attention to its presentation in a late childhood.

Keywords: Pulmonary atresia; anomalous origin; right sided aortic arch.

1. INTRODUCTION

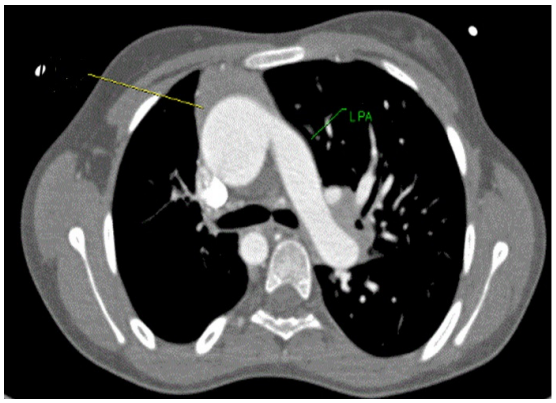
Unilateral absence of pulmonary artery or agenesis is a rare congenital disorder with a prevalance of about 1/200000. It was reported for the first time in 1868 by Fraentzel [1]. Usually it is accompanied by cardiovascular malformations some of which may detoriorate the clinical status of the patient such as coarctation of aorta and transposition of great arteries [2]. It is two times more frequent in the right side. Although left sided agenesis is associated with life threatening malformations which requires early diagnosis and surgical repair, patients with right sided agenesis may survive asyptomatic until aduldhood [3,4].

Anomalous origin of the pulmonary artery (AOPA) from the aorta is a rare congenital malformation. It is embryologically associated with a defect in the involution of the proximal sixth aortic arch and fifth aortic arches in the affected side [5]. Right sided anomalous origin is seen five times more than the left sided one [6,7]. AOLPA is mostly associated with tetralogy of Fallot with a ratio of 75% [6].

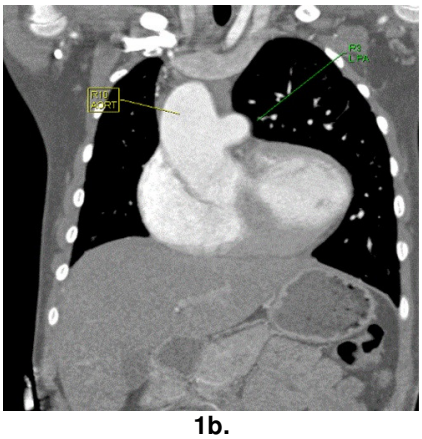
In this report we presented the computed tomography angiography findings of an incidentally detected case of anomalous origin of the left pulmonary artery from the aorta with a right sided aortic arch associated with pulmonary atresia without central pulmonary artery supplying the right lung.

2. CASE

13 year-old girl was referred to our pulmonary clinic because of cough, dyspnea and fever. Our patient reported frequent respiratory infections and exertional dyspnea. There was no medication history. On her physical examination there were rales over the apical region of the left lung. On her chest X-ray there was consolidation on left upper lobe. But also cardiothoracic ratio was slightly increased and the shadow of the arch aorta wasn't seen on the left. So echocardiography was performed and on assessment pulmonary arteries couldn't be visualized. Computed tomography angiography (CTA) was performed to visualize the anatomy. On her CTA a right-sided aortic arch, with absence of the pulmonary artery was detected. Left pulmonary artery was originating from the ascending aorta just above the sino-tubular junction Fig. 1a, 1b, 3. There were



1a.



1b.

Fig. 1a,b. Axial (1a) and coronal (1b) maximum intensity projection (MIP) images showing the anomalous origin of the left pulmonary from the ascending aorta just above the sinotubular junction. Also the right-sided arcus is seen

well-developed collateral blood supply to the right lung Fig. 2. Also there was associated consolidation in the apicoposterior segment of the left upper lobe Fig. 4. Her pulmonary artery tension values were in normal limits. Because of her clinical symptoms relating the pneumonia she was given antibiotherapy. Surgery was not performed to the patient and she was conservatively treated for her cardiovascular status.

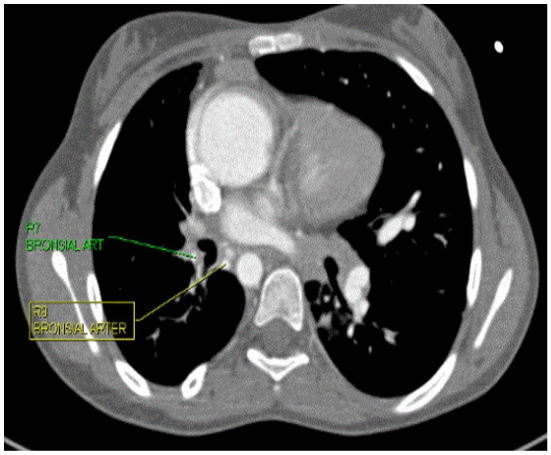


Fig. 2. Well developed bronchial arteries supplying the right lung are seen on axial MIP images

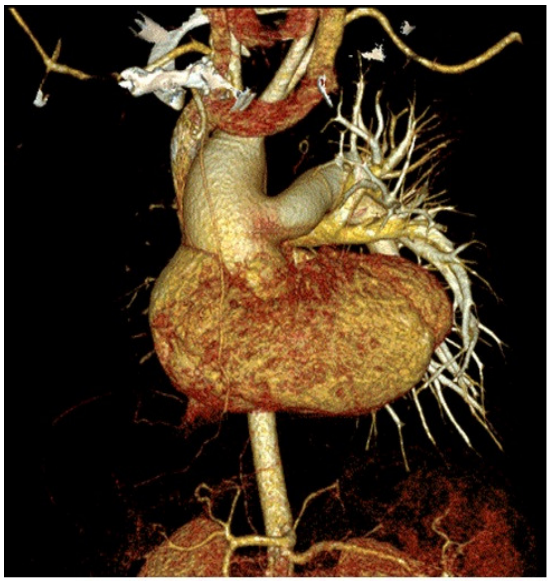


Fig. 3. Volume rendered (VR) images of the same patient; showing the originating left pulmonary artery from ascending aorta

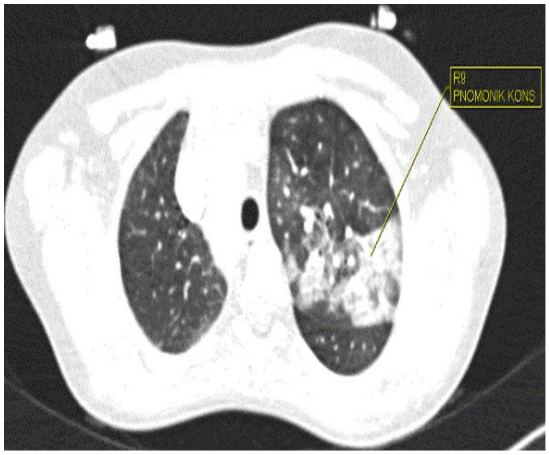


Fig. 4. Axial images in the parenchymal window shows the consolidation on the left upper lung

3. DISCUSSION

The anomalous origin of one pulmonary artery from the aorta is a very rare entity. In most cases anomalous origin of the right pulmonary artery is seen but as in our case the anomalous origin of the left pulmonary artery from the aorta is an extremely rare entitiy [8]. The underlying pathogenesis is a developmental abnormality of the aortic and branchial arches. While failure in resorption of the fifth branchial arch with an existing sixth branchial arch leads to anomalous origin of the right pulmonary artery, failure in the development of fifth and sixth branchial arches results in anomalous origin of the left pulmonary artery from the aorta as in our case. [5]. Beside its association with cardiovascular malformations; mostly tetralogy of Fallot, AOLPA is usually associated with right sided aortic arch. In our case there was no associated tetralogy of Fallot and the coexisting cardiovascular anomalies were atresia of the right pulmonary artery and right sided archus aorta [2].

The clinical presentation includes heart murmurs, tachypnea, dyspnea, heart failure and recurrent respiratory infections in early years of life [9,10]. Our case was presented with recurrent pulmonary infections and sypmtoms of heart failure such as exertional dyspnea but with a relatively late onset of sypmtoms probably associated with the well developed collateral blood supply to the right lung and absence of life-threatening coexisting abnormalities such as coartation of aorta or tetralogy of Fallot.

The diagnosis of AOPA can be achieved by echocardiography, cineangiography, magnetic resonance imaging and computed tomography angiography [11,12]. Computed tomography angiography is useful in the diagnosis with its high temporal and spatial resolution. It allows also a three dimentional view and visualization of the complex vascular anatomy. In our case the left pulmonary artery couldn't be visualized in echocardiography and by computed tomography angiography both the anatomy of the pulmonary arteries and the coexisting infiltration in the left lung detected at the same time.

4. CONCLUSION

The anomalous origin of pulmonary artery from the ascending aorta especially the left pulmonary artery is a rare anomaly with some reported coexisting abnormalities such as tetralogy of Fallot mostly presenting in the early years of life. Different imaging modalities both noninvasive and invasive can be used for the diagnosis.

We have reported with this case an anomalous origin of the left pulmonary artery from ascending aorta with coexisting right sided aortic arch and right pulmonary atresia presenting with recurrent pulmonary infection and diagnosed incidentally. And we wanted to take attention to its presentation at a relatively late age and diagnosed noninvasively by computed tomography angiography.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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