CASE REPORT

AGENESIS OF LEFT PULMONARY ARTERY WITH RIGHT SIDED AORTIC ARCH IN ADULT FEMALE PATIENT: A CASE REPORT

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ABSTRACT

OBJECTIVE: Unilateral pulmonary artery agenesis (UPAA) is a rare congenital anomaly due to a malformation of the sixth aortic arch of the affected side during embryogenesis. The diagnosis is usually set at adolescence, however it can remain asymptomatic and late diagnosis is possible. We discuss role of cross sectional imaging of asymptomatic adult female patient with pulmonary artery agenesis. **RESULT:** We present a case of 66 years old female who was known case of pancreatic adenocarcinoma however her CT chest was done as part of staging protocol, which showed left pulmonary artery agenesis with right sided aortic arch. **CONCLUSION:** Contrast-enhanced CT is non-invasive and adequate imaging modality for diagnosis of UPAA. It can also depict associated aortic arch and congenital cardiac anomalies and help in assessing PH which can be seen in such patients. Though UPAA is rare anomaly but it should be included in the differential diagnosis of a chest radiograph with a small hyper-transradiant hemithorax

Key words: Pulmonary artery, agenesis, congenital

Introduction

Congenital unilateral pulmonary agenesis (UPAA) and hypoplasia of a pulmonary artery (UHPA) are rare congenital anomalies.[1]

Unilateral agenesis of pulmonary artery is commonly associated with congenital cardiovascular anomalies; However less frequently, It may be seen as an isolated abnormality. Majority of the cases reported in the literature, shows that patients ususally presents early and are diagnosed in infancy or childhood however few patients appear to remain asymptomatic until adulthood.[2]

UPAA can be accompanied by other congenital heart abnormalities, but this was not observed in our patient. In almost 2% of the cases, absent pulmonary artery and the aortic arch are at the same side.[3,4]

We present a case of 66 years old female who underwent contrast enhanced CT as a part of her staging workup for pancreatic head adenocarcinoma.

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Case Discussion

A 66 years old female patient resident of Lahore presented with weight loss and jaundice. She was known case of diabetes and hypertension. Was treated for pulmonary tuberculosis in past. Her base line investigations showed deranged LFTs with raised bilirubin and alkaline phosphatse however CA 19.9 was raised to 2400 U/ml. Imaging showed a pancreatic head mass which was resectable and stent was passed for distal CBD stricture. She further underwent a x-ray chest and Contrast enhanced CT chest on canon 160 slicer scanner for staging workup, which was negative for pulmonary metastasis. X-ray showed deviation of the mediastinum to the left side obscuring details of the ipsilateral lung with reticular changes and volume loss suggestive of underlying fibrotic changes (Fig. 1).

CT chest showed agenesis of left pulmonary artery with multiple collateral circulation seen in mediastinum

and left-sided intercostal spaces (Fig. 2a). Hypoplastic left lung with ipsilateral shift of heart and mediastinum was noted. Compensatory hyperinflation of the right lung was seen. Right-sided aortic arch was also noted (Fig. 2b). Fibrotic changes were seen in both lungs, more marked on the left side.

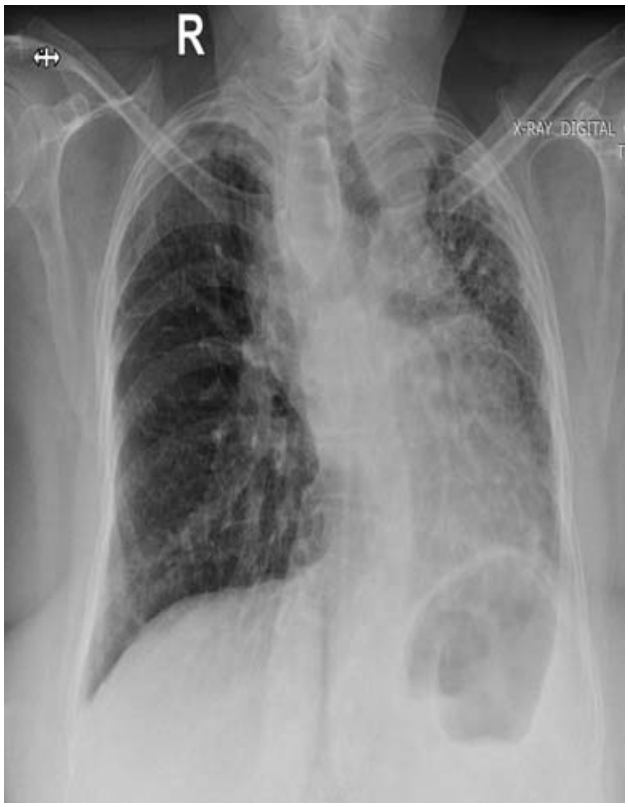
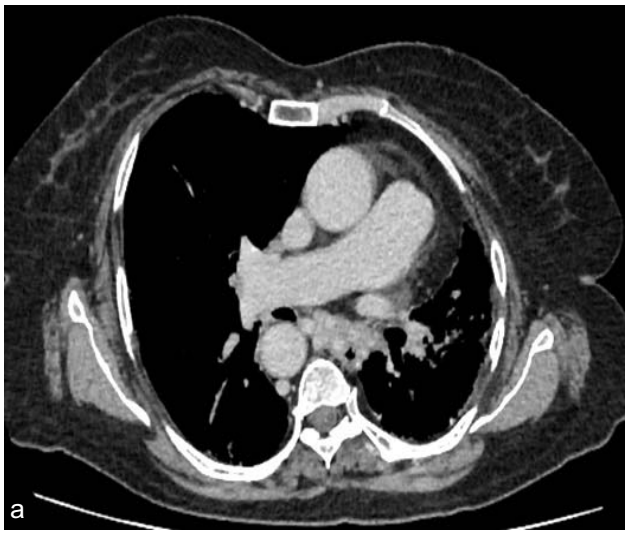


Figure 1: Xray chest PA view: left sided volume loss with reticular opacities. Ipsilateral medistainal shift and hyperinflated right lung.



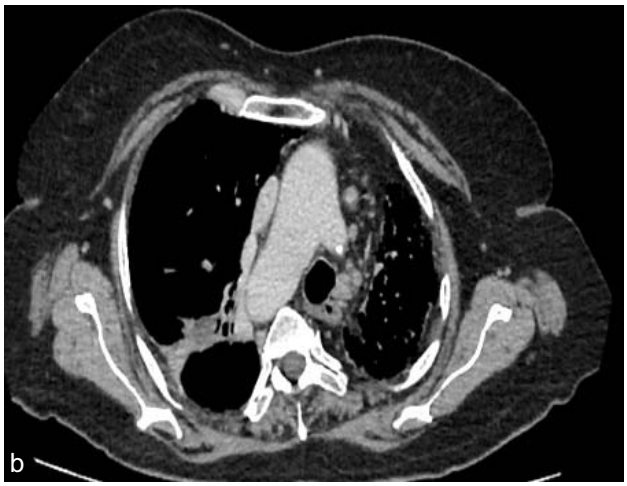


Figure 2a,b: CE CT chest axial images: showing left pulmonary artery agenesis with right sided aortic arch.

Discussion

UPAA was first diagnosed in 1868[5] and since then, 420 cases have been described in the literature. The estimated prevalence of single UPAA is 1/200,000 patients, and there is no sex predilection. While average age of diagnosis is 14 years, in our case the diagnosis was set in adulthood. Despite the fact that in 2/3 of cases in which right side is affected, our patients had a left side UPAA.[3]

Patients with UPAA have a normal pulmonary trunk and unilateral absence of a pulmonary artery branch. Intrapulmonary vasculature and the distal part of the pulmonary trunk may develop normally and receive blood supply from the bronchial vessels,[6] resulting in a small, hypovascular lung at the affected side.

UPAA can be accompanied by other congenital heart abnormalities1 but this was not observed in any of our patient. In almost 2% of the cases, the aortic arch is at the same side of the absent pulmonary artery,[7] however ever in our case aortic arch was on right and agenesis of pulmonary artery was observed on left. Blood in the affected lung is supplied by the bronchial arteries or by abnormal collaterals arising from the bronchial, subclavian, intercostal and sub-diaphragmatic arteries.[1,7] Our patient had multiple collateral vessels in the mediastinum and intercostal region.

Symptoms may include dyspnea on exertion, recurrent pulmonary infections, hemoptysis (in 20% of patients), chest pain, or pleural effusion. Massive, life-threatening

hemoptysis could also occur. Pulmonary hypertension(PH) is observed in 25% of the patients and determines long-term survival.[1,7] According to study of Bouros et al[5] who described 6 cases of isolated pulmonary artery agenesis in adulthood. Only 1 case was asymptomatic, while the others had recurrent respiratory tract infections. Additionally, 1 patient had hemoptysis and dyspnea.[8] In our case patient had history of treated pulmonary tuberculosis but had no pulmonary symptoms in childhood or currently.

Findings at the chest X-ray may include cardiomediastinal displacement, absence of the pulmonary artery shadow on the affected side, an ipsilateral elevation of the diaphragm, a contralateral compensatory hyperinflation of the hemithorax.[8]

CT is adequate for UPAA diagnosis, limiting the use of invasive techniques. Parenchymal findings can also be well depicted on CT.[1] In addition, chest CT provides useful information regarding PH[9] or congenital heart defects.

Conclusion

Contrast-enhanced CT is non-invasive and adequate imaging modality for diagnosis of UPAA. It can also depict associated aortic arch and congenital cardiac anomalies and help in assessing PH which can be seen in such patients. Though UPAA is rare anomaly but it should be included in the differential diagnosis of a chest radiograph with a small hyper-transradiant hemithorax

Conflict of Interest: None

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