Original Research Paper

Volume - 12 | Issue - 12 | December - 2022 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Pulmonary Medicine

E - POSTER : ABSENT RIGHT PULMONARY ARTERY WITH PATENT DUCTUS ARTERIOSUS

Dr. Arawinth Raj	Final Year Resident
Dr. Sajith	Associate Professor
Dr. Dilip S Phansalkar	Professor & HOD

KEYWORDS :

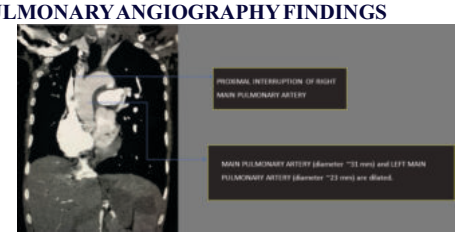
INTRODUCTION

- Unilateral absence of pulmonary artery (UAPA) is a rare congenital anomaly which was first described by Fraentzel in 1868 .
- It is frequently associated with other cardio-vascular anomalies, such as Fallot of tetralogy, septal defects and patent ductus arteriosus.
- We present a case of absent right pulmonary artery with patent ductus arteriosus in a 42 years old female patient with findings in CT pulmonary angiography.

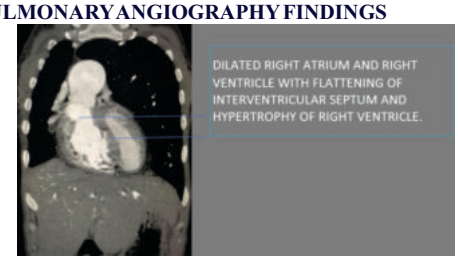
CLINICAL INFORMATION

- Our patient was a 42 years old female with complaints of breathlessness over the past 6 months.
- She also had history of cough with mucoid expocteration for the past 3 months.
- She is a known case of pulmonary Tuberculosis (6 years ago)and had taken ATT for 6 months.
- On examination, patient was febrile (Temp - 99 degrees) and she had pan digital clubbing (grade III).
- Blood work up showed leucytosis , elevated infective markers , elevated lactates and hyponatremia.
- CVS examination showed - S1, S2 normal and no murmur.• Echocardiography was done and revealed right atrium and right ventricular dilatation.
- CT pulmonary angiography was proceeded to look for pulmonary embolism.

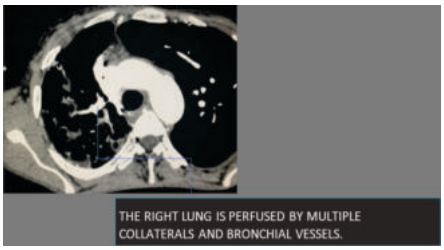
CT PULMONARY ANGIOGRAPHY FINDINGS



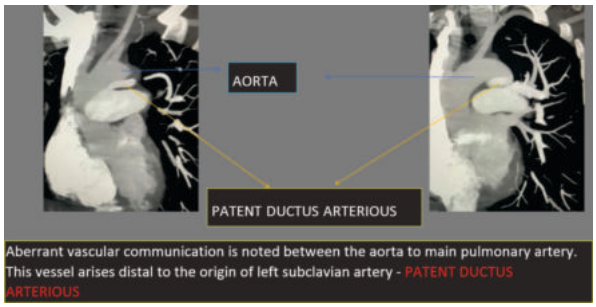
CT PULMONARY ANGIOGRAPHY FINDINGS



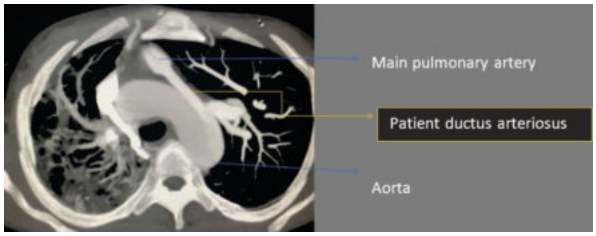
CT PULMONARY ANGIOGRAPHY FINDINGS

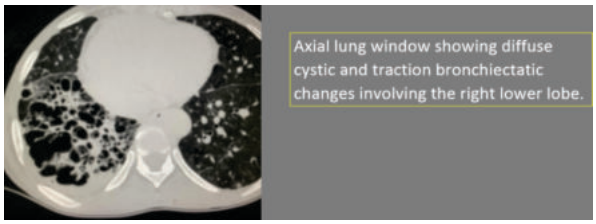


CT PULMONARY ANGIOGRAPHY FINDINGS



CT PULMONARY ANGIOGRAPHY FINDINGS





DIAGNOSIS

- Unilateral Absent Right Pulmonary Artery With Patent Ductus Arteriosus.
- Pulmonary Hypertension.
- Bronchiectatic Changes In Right Lung .

DISCUSSION

- The exact embryological cause of UAPA is a matter of debate usually secondary to altered development of the six aortic arch segment.
- The intrapulmonary pulmonary arteries arise from the lung buds and the extrapulmonary pulmonary arteries arise from the proximal portion of the sixth aortic arch.
- The main PA is derived from the truncoaortic sac.
- The ductus arteriosus, which forms from the distal portion of the sixth arches, connects to the primitive dorsal aorta, which becomes the underside of the aortic arch ipsilateral to the arch or the base of the innominate artery contralateral to the arch.
- An absent PA is caused by the involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary PA to the distal sixth aortic arch.
- The distal intra-pulmonary branches of the affected artery remain intact and can be supplied by collateral vessels from bronchial, intercostals, internal mammary, subclavian or even coronary arteries .

- Many patients with isolated UAPA may remain asymptomatic until they reach adulthood while others may complain of recurrent respiratory tract infections, dyspnoea on exertion, pulmonary oedema, pulmonary hypertension or hemoptysis in contralateral lung.
- Pulmonary hypertension occurring as a sequel to UAPA may result from an increase in blood flow to the contralateral unaffected artery. This leads to chronic vasoconstriction of the pulmonary arteries with subsequent pulmonary hypertension.
- The aetiology of recurrent infection may be due to decrease the blood flow to the affected lung with subsequent poor delivery of the inflammatory cells associated with alveolar hypocapnia leading to bronchoconstriction and mucous trapping .
- Hemoptysis may occur due to the large collateral circulation .
- In our case, the patient had multiple episodes of respiratory tract infections. Axial lung window showed cystic and bronchiectatic changes in right lung field sequelae to previous insult. Echocardiography showed right atrium and right ventricle dilatation and CT pulmonary angiography was done to rule out pulmonary artery hypertension.
- Cardiac Enlargement: Cardiac enlargement was present in 73 per cent of the cases. In more than half of these the enlargement was either moderate or marked in degree.
- Treatment options for these patients include partial or total pneumonectomy, closure of selected collateral arteries not solely responsible for pulmonary blood flow or a primary versus staged pulmonary artery anastomosis.

CONCLUSION

- An absent pulmonary artery is rare and may escape clinical detection even into a patient's adulthood if the condition is unilateral and isolated .
- Radiologists should be aware of their potential role in early detection of this entity; even relatively asymptomatic patients with a unilateral absent pulmonary artery may have significant morbidity.

REFERENCES

1. Keats, Theodore E., and Howard L. Steinbach. "Patent Ductus Arteriosus." Radiology, vol. 64, no. 4, Radiological Society of North America (RSNA), Apr. 1955, pp. 528–37. Crossref, https://doi.org/10.1148/64.4.528.
2. Apostolopoulou, Sotiria C., et al. "'Absent' Pulmonary Artery in One Adult and Five Pediatric Patients: Imaging, Embryology, and Therapeutic Implications." American Journal of Roentgenology, vol. 179, no. 5, American Roentgen Ray Society, Nov. 2002, pp. 1253–60. Crossref, https://doi.org/10.2214/ajr.179.5.1791253
3. Kruzliak, Peter, et al. "Unilateral Absence of Pulmonary Artery: Pathophysiology, Symptoms, Diagnosis and Current Treatment." Archives of Cardiovascular Diseases, vol. 106, no. 8–9, Elsevier BV, Aug. 2013, pp. 448–54. Crossref, https://doi.org/10.1016/j.acvd.2013.05.004.
4. SWAN, HENRY. "Absent Left Pulmonary Artery and Right-Sided Patent Ductus Arteriosus." Archives of Surgery, vol. 87, no. 2, American Medical Association (AMA), Aug. 1963, p. 196. Crossref, https://doi.org/10.1001/archsurg.1963.01310140004003
5. Lohchab, Shamsher Singh, and Kuldeep Laller. "Large Tortuous Patent Ductus Arteriosus With Proximal Left Pulmonary Artery Hypoplasia." World Journal for Pediatric and Congenital Heart Surgery, vol. 2, no. 3, SAGE Publications, July 2011, pp. 526–27. Crossref, https://doi.org/10.1177/2150135111403782.