Case Report

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Incidentally Detected Isolated Unilateral Pulmonary Artery Agenesis

Radiology Section

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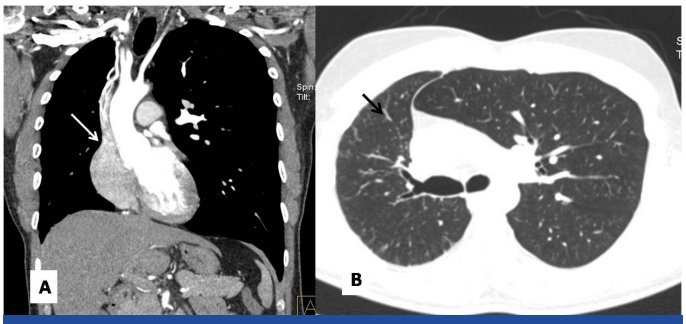
ABSTRACT

We are reporting here, a case of an incidentally detected, isolated, unilateral pulmonary artery agenesis on contrast enhanced Computed Tomography (CT) of the chest in a 39-year-old male patient.

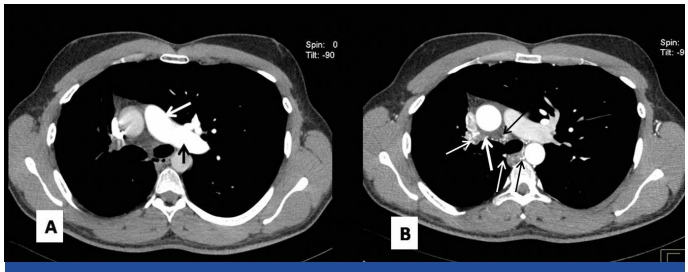
Key Words: Pulmonary artery, Agenesis, Isolated

CASE REPORT

A 39-Year-old male patient who had come for a routine health check up was found to have a reduced right lung volume on the chest X-Ray. The patient was subjected to a contrast enhanced Computed Tomography (CT) scan of the chest on a Siemens somatom emotion 6CT scanner, with acquisition from the diaphragm to the lung apex after injecting an intravenous contrast. The CT Scan showed a reduced right lung volume with mediastinal and cardiac shifts to the right side [Table/Fig-1(A) and 1(B)]. The contra lateral lung showed hyperinflation. There was a complete absence of the right pulmonary artery with normal main and left pulmonary arteries [Table/Fig-2(A)]. The right lung was supplied by the systemic arteries [Table/Fig-2(B)] which arose from the descending thoracic, the suprarenal abdominal aorta and the proximal left subclavian arteries. The venous drainage of the right lung was into the pulmonary veins, which drained into the left atrium. An echocardiogram which was done, did not reveal any evidence of congenital cardiovascular defects or pulmonary hypertension.



[Table/Fig-1]: (A) Coronal multiplanar reformatted CT image showing cardio-mediastinal shift to right side (white arrow). (B) Axial CT image in lung window section shows hypoplastic right lung (black arrow).



[Table/Fig-2]: (A) Axial Contrast CT image showing normal main (white arrow) and left pulmonary artery (black arrow) with absent right pulmonary artery. (B) Axial Contrast CT image showing multiple collaterals from aorta supplying right lung (white and black arrows).

DISCUSSION

Unilateral Pulmonary Artery Agenesis (UPAA) is a rare anomaly and it is commonly associated with congenital cardiovascular defects such as the Fallot tetralogy and septal defects. The prevalence of this anomaly is estimated to be 1 in 300,000 [1]. The other cardiovascular anomalies which are associated with unilateral pulmonary artery agenesis include coarctation of the aorta, patent ductus arteriosus, subvascular aortic stenosis and transposition of the great arteries. Usually, in these conditions, the left pulmonary artery is more commonly absent and the patients present in infancy or in early childhood [1]. Isolated UPAA is very rare and it is not detected until adult hood, like in our case. Isolated UPAA commonly affects the right pulmonary artery [2]. The embryologic reason for an absent pulmonary artery is involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary pulmonary artery to the distal sixth aortic arch. The extrapulmonary arteries arise from the proximal portion of the sixth aortic arch and the ductus arteriosus forms from the distal sixth aortic arch [1]. These patients usually present with symptoms like recurrent respiratory tract infections, dyspnoea and haemoptysis [1,3]. Few of them may present with pulmonary artery hypertension, pulmonary haemorrhage, high altitude pulmonary oedema or heart failure [1]. There is no consensus for treating isolated UPAA. While there are those who believe in treating only symptomatic patients, others advocate a staged repair to restore the physiological pulmonary circulation [4].

REFERENCES

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