Congenital isolated unilateral hypoplasia of the left pulmonary artery: A rare incidental anomaly in an elderly female

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ABSTRACT

Congenital isolated unilateral hypoplasia of the pulmonary artery (CIUPAH) can have late presentation in the adulthood, mostly diagnosed incidentally, as in our case. They may have symptoms such as exertional dyspnea, recurrent lung infections, and hemoptysis. Although the surgical management is a definitive treatment modality, it depends on the presence of reversible pulmonary hypertension and size of the pulmonary arteries; interventional and medical management can be attempted as a palliative therapy in cases where definitive surgical management is not feasible. We have discussed a rare case of isolated unilateral pulmonary artery hypoplasia which is a congenital anomaly of the lung vasculature without associated congenital heart disease manifested in adulthood.

KEY WORDS: Hypoplastic lung, hypoxia, pulmonary hypertension, respiratory distress

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A 65-year-old female presented with fever and breathlessness for 4 days. There were no recurrent pulmonary infections in the past. Vital parameters recorded were a pulse rate of 122/min, a blood pressure of 110/70 mmHg, respiratory rate of 38/min, body temperature 40.1°C, and room air saturation was 78%. On auscultation, there was decreased air entry to the right lower lobe with coarse crepitation. Arterial blood gas analysis showed Type II respiratory failure. A team of emergency physicians ventilated her, which improved saturation to 95%.

Laboratory parameters revealed elevated total leukocyte count and acute-phase reactants with neutrophilic predominance. Chest X-ray revealed consolidation of the right lower and middle lobe, hypoplasia of the left lung, elevated left hemidiaphragm, and shift of the mediastinum to the left with prominent right pulmonary artery shadow. Echocardiography showed mild dilatation of the right atrium and ventricle with right ventricular systolic pressure of 55 mmHg. Computed tomography (CT) showed a hypoplastic left lung along with left pulmonary artery hypoplasia. There was reduced vascularity of the left lung. The right pulmonary artery was dilated [Figure 1a-d]. The patient was discharged after 21 days of hospitalization.

Congenital isolated unilateral hypoplasia of the pulmonary artery (CIUPAH) or agenesis is a rare congenital anomaly without any coexisting congenital cardiovascular anomalies and should be considered as a clinical continuum.[1]

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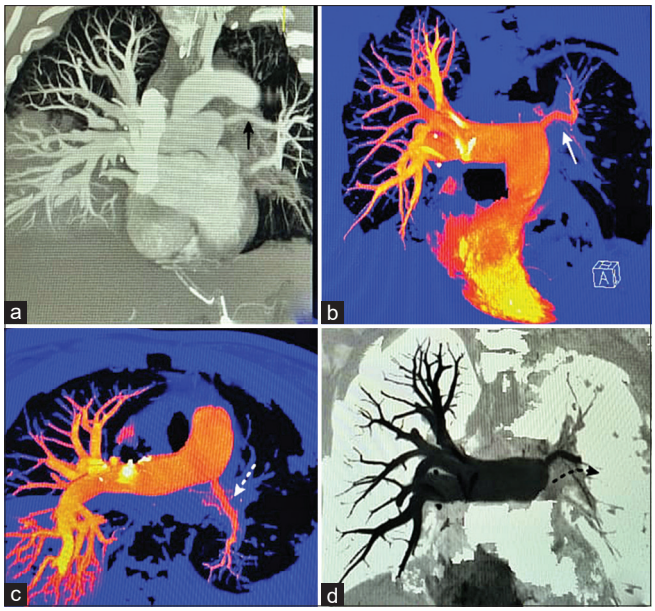

Figure 1: (a-d) Computed tomography angiography of the pulmonary artery. Coronal maximum intensity projection (MIP) image at the level of the main pulmonary artery bifurcation shows hypoplastic left pulmonary artery (black solid arrow, Panel A) with homogeneous opacity of the right lower lobe suggestive of consolidation. Coronal selective color-coded MIP image at the level of main pulmonary artery bifurcation shows hypoplastic left pulmonary artery (solid white arrow, Panel B). Axial selective color-coded MIP image at the level of main pulmonary artery bifurcation shows normal-sized normal, right pulmonary artery and hypoplastic left pulmonary artery (dashed white arrow, Panel C). Axial invert gray scale MIP image at the level of main pulmonary artery showed hypoplastic left pulmonary artery (dashed black arrow, Panel D)

Nitta *et al*. documented a case of left pulmonary artery hypoplasia with left gastric artery pseudoaneurysm ruptured with a posterior mediastinal hematoma. They summarized 11 previous CIUPAH cases published in the literature since 1958 with those presenting at adulthood (>19 years) and the eldest patient being 65 years old.[2] Chang *et al*. identified a case of CIUPAH in which a 68-year-old male with a retrograde flow from the right pulmonary hypoplastic artery to the left dilated pulmonary artery due to systemic feeding was collateral to the right pulmonary hypoplastic artery.[3]

Park *et al*. reported a case of a 36-year-male who had recurrent hemoptysis, and CT of the chest revealed left pulmonary hypoplasia with increased mediastinum vasculature and old pulmonary infarctions with pulmonary bands and linear opacities.[1]

The above two cases were not included in the series of 11 cases of CIUPAH by Nitta *et al*. So far, the literature describes 14 cases of congenital CIUPAH in the adult age group, including our case.

CIUPAH is a rare congenital anomaly in adults compared to an isolated unilateral agenesis of the pulmonary artery, and its survival until the sixth decade is relatively uncommon. This condition should be suspected if the vascular discrepancy between the two lungs is present.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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