

Unilateral Pulmonary Fibrosis with Pulmonary Artery Agenesis (PAA): A Case Report

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Abstract: Unilateral pulmonary artery agenesis (AUAP) is a rare clinical condition that can be seen either in isolation or in combination with other congenital vascular disorders. When isolated, the patient may be almost asymptomatic and only become aware of this anomaly through medical examinations performed for other reasons.

Here we discuss the case of a female patient with chest CT evidence of unilateral fibrous thickening, with pulmonary hypoexpansion; investigations revealed the coexistence of AUAP.

This clinical association is described by very rare case-reports in the literature.

Keywords: Interstitial lung disease, Unilateral pulmonary fibrosis, Pulmonary artery agenesis.

INTRODUCTION

Pulmonary interstitial diseases are a heterogeneous group of pathologies affecting the pulmonary interstitium, a field of respiratory diseases that is not so much dealt with by the territorial medicine and that is more pertinent to few hospital reference centers.

By definition, pulmonary interstitial diseases affect both parenchymal areas, being diffuse infiltrative diseases. The cases of unilateral pulmonary fibrosis represent a rarity often secondary to other diseases such as autoimmune diseases or vascular abnormalities [1], such as pulmonary artery agenesis (PAA), a rare condition caused by developmental abnormalities of the ipsilateral sixth aortic arch resulting in a deficit of blood supply to one lung, which is perfused by the collateral circulation.

Herein we report a case of a female patient with a interstitial lung disease at chest CT scan with a coexisting pulmonary artery agenesis.

Case Presentation

A 55-year-old female presented to my observation in September 2021 with a long history of dry cough, and breathlessness; she was a former homemaker who smoked about 20 cigarettes a day, in addition to passive exposure to cigarette smoke for more than 20 years.

She was in Inhalation Therapy with inhaled corticosteroids with broncodilators for a history of Asthma; a previous spirometry performed in 2020 reported a non-reversible bronchial obstruction with FEV1/FVC <70%, FEV1 65 % of predicted, FVC

3.50 Liters (75% of the predicted). An additional comorbidity was hypertension in treatment and under cardiological monitoring.

A previous chest X- ray showed a mild reinforcement of the bronco-vascular pattern, most noticeable in the left lung.

Investigations

On examination, her respiratory rate was 20 breaths per minute with heart rate of 75 and oxygen saturation of 95% at room temperature. Her blood pressure was 120/75 mmHg.

There were fixed crepitations (“velcro-rales” like) on chest auscultation, not modifiable by coughing, in the left field.

A routine blood tests as not detected significant anomalies.

A chest CT scan showed modest and widespread interstitial thickening on the left with a right aortic arch and absence of definite lesions of the lung parenchyma or pleural spaces (Figure 1) and complete sparing of right lung.

In view of the chest CT scan performed, and of the diagnostic suspicion, it was decided to perform a chest Angio-CT evaluation that the patient performed only a few months later, with evidence of agenesis of the left

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Figure 1: CT scan of the chest with interstitial thickening of the left lung.

pulmonary artery and evidence of multiple tributary collateral circles of the left subclavian, internal mammary and peribronchial aorta originating from the descending aorta, which run along the left mediastinal margin. The pulmonary veins were also reduced in caliber compared to control, and the left lung was hypoexpanded (Figure 2) and affected by thickening of the axial and peripheral interstitium with thickening of the peribronchial septa and fibrotic striae.

The pulmonary trunk and right pulmonary artery were regularly opacified with compensatory overexpansion of the right lung. This was also associated with right-posterior aortic arch with left common trunk.



Figure 2: CT scan of the chest with left lung hypoexpansion and interstitial thickening.

A cardiological evaluation with echocardiography was then requested, which excluded major cardiac functional problems (ejection fraction: 60%) and/or valvular abnormalities.

A vascular surgery consultation did not detect abnormalities in vascular flows with preserved blood distribution, and indicated periodic follow-up.

The patient also underwent diffusion lung CO test, which resulted in a 65% reduction.

Bronchial obstruction was also confirmed with indication for continuation of the current inhalation therapy (inhaled corticosteroid and long-acting beta2 agonist).

DISCUSSION

Unilateral absence of the pulmonary artery (AUAP) is a rare congenital anomaly that can occur in isolation or in association with other cardiovascular anomalies (tetralogy of Fallot, interatrial and interventricular septal defect, aortic coarctation, truncus arteriosus, and pulmonary valve atresia); a right-posterior aortic arch is also possible, as for our patient.

A complete absence is caused by involution of the proximal sixth aortic arch, with failure to connect with the pulmonary trunk.

AUAP in its isolated form is rare, and patients without associated cardiac abnormalities (as our patient) usually grow out of it without developing significant symptoms. When diagnosed in adulthood, this condition is often incidentally detected during chest computed tomography performed for other reasons, and thus knowledge of this condition is also important for radiologists.

Although there are several treatment options including surgery, endovascular embolization, a conservative management is often preferred, also in view of the fact that there is actually no broad consensus on the optimal treatment.

Unilateral pulmonary fibrosis is also a rare diagnosis; possible causes include gastroesophageal reflux or autoimmune diseases [1], pulmonary damage from radiation or mechanical ventilation [2], chronic local parenchymal inflammation, or vascular abnormalities as in the case discussed [3].

The association between agenesis of a pulmonary artery branch (AUAP) and unilateral pulmonary fibrosis represents a particular rarity, although already mentioned in the literature by few case-reports [4].

This rare clinical case should give pause for thought about the need for a better and broader classification of interstitial lung diseases, as they are well known and widely characterized only for the classic bilateral forms, which are markedly more frequent, and little known in the rare unilateral forms.

CONCLUSIONS

Unilateral pulmonary fibrosis is a diagnosis of particular difficulty for Pulmonologists and Radiologists because of its particular rarity, especially when associated with congenital vascular anomalies as in the case described.

It is important to increase the knowledge of this pathological entity with its possible associations to improve the early diagnosis of treatable cases.

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AUTHORS' CONTRIBUTION

Conceptualization and writing: *Gianfranco Scotto di Frega*.

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