

## Case Report

# A rare case of underlying pulmonary sequestration in a patient with recently diagnosed medium and large vessel vasculitis

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## ABSTRACT

Vasculitis of medium- and large-sized arteries is an inflammatory and stenotic disease characterized by a strong predilection for the aortic arch and its branches. It presents with symptoms and signs as per the vessels and organs involved. Pulmonary sequestration is a rare abnormality characterized by a mass of nonfunctioning lung tissue that receives its vascular supply from a systemic artery and is separated from the normal tracheobronchial tree. The following is a rare case report showing the presence of pulmonary sequestration in a patient with recently diagnosed hypertension and intestinal angina due to medium and large vessel vasculitis.

**KEY WORDS:** Intralobar lung sequestration, lung sequestration, vasculitis

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## INTRODUCTION

Vasculitis of medium- and large-sized arteries is an uncommon disease with an estimated annual incidence rate of 1.2-2.6 cases per million.<sup>[1]</sup> Vasculitis may present with various local and systemic clinical features. Local symptoms pertain to the artery involved. Systemic symptoms may manifest in the form of fever, arthralgia, night sweats and weight loss. Pulmonary sequestration consists of a nonfunctioning mass of normal lung tissue that lacks normal communication with the tracheobronchial tree, and that receives its arterial blood supply from the systemic circulation. It is estimated to comprise 0.15 to 6.4 percent of all congenital pulmonary malformations, making it an extremely rare disorder.

Pulmonary sequestration is a rare manifestation associated with medium and large vessel vasculitis. We describe such a rare case.

## CASE REPORT

A 25-year-old female presented with complaints of malaise, weight loss, and diffuse abdominal pain, more in the umbilical region since 1-2 months. Pain was aggravated with meals and relieved with fasting. On examination, her blood pressure was 160/92 mm Hg in right arm and 158/90 in left arm; lower limb blood pressure was 126/80 in right lower limb and 128/82 in left lower limb. Pulse rate was 90 per minute, regular, good volume with no radiofemoral delay. A renal bruit was heard on the left side. Rest of the systemic examination was within normal limits. Complete blood count, liver function tests, kidney function tests, and urine routine microscopy were within normal limits. Erythrocyte sedimentation rate (ESR) was raised to 35 mm/hour and C-reactive protein (CRP) was more than 5 mg/L. Her chest X-ray and electrocardiogram (ECG) were normal. Her fundus examination was normal. Serology for HIV 1 and 2, hepatitis B surface antigen (HBsAg), anti hepatitis C virus (anti-HCV), venereal disease research laboratory (VDRL) test, anti-nuclear antibodies (ANA), cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) and perinuclear antineutrophil cytoplasmic antibodies (p-ANCA) were negative. Complement levels were normal. Ultrasonography (USG) of the abdomen was normal. Contrast enhanced computed tomography (CT) scan of the thorax and abdomen was done, which revealed thickening of the descending aorta upto bilateral renal artery origin with thickening of the

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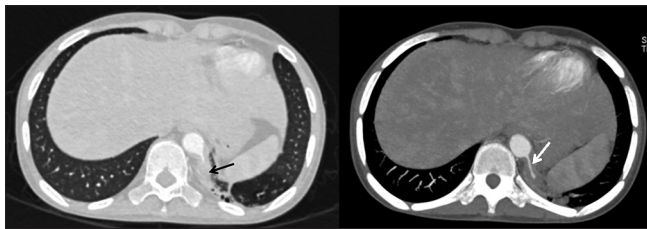
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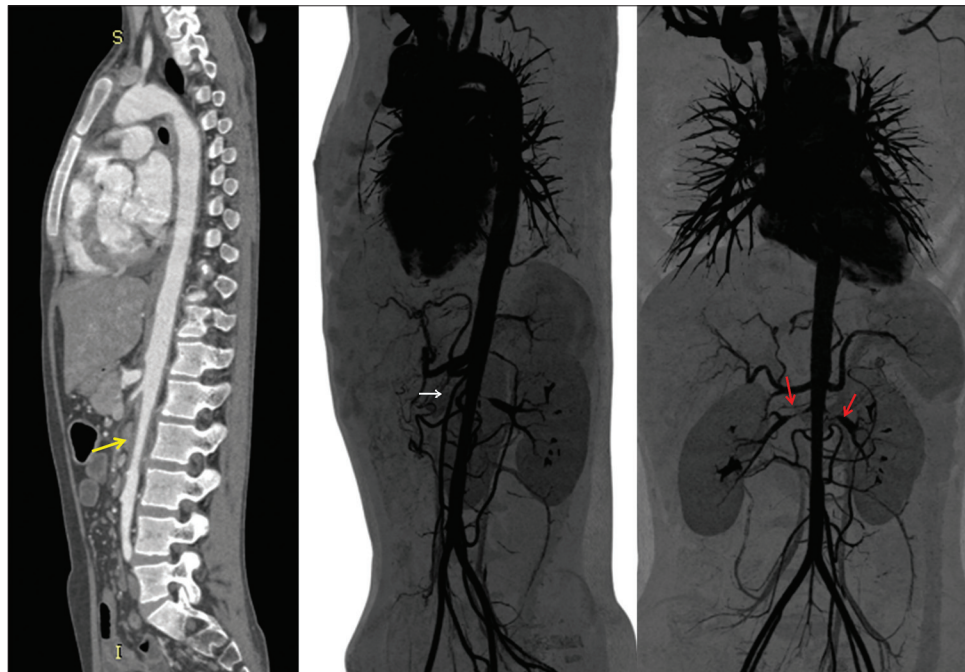
superior mesenteric artery. CT angiography was done, and it revealed wall thickening of the descending aorta with narrowing of lumen. There was also thickening of the superior mesenteric artery with narrowing of lumen. Pulmonary trunk and bilateral pulmonary arteries were also normal. There was evidence of a heterogeneous, hyperdense region in the left medial basal segment deriving its arterial supply separately from the aorta. There was no direct communication with the tracheobronchial tree, and the heterogeneous lung segment had no separate pleural covering. The above-mentioned findings were suggestive of an intralobar pulmonary sequestration [Figure 1]. Bilateral renal artery showed stenosis with involvement of the ostia and proximal segment with post-stenotic dilatation [Figure 2]. The other branches of the abdominal aorta were normal. The thoracic aorta and its branches were also normal in course and caliber. The findings were consistent with large vessel vasculitis.



**Figure 1:** Axial computed tomography sections of the thorax showing heterogeneous segment (black arrow) in the medial basal segment of the left lung deriving its arterial supply from the descending aorta (white arrow)

## DISCUSSION

Vasculitis of medium- and large-sized arteries is an uncommon disease with an estimated annual incidence rate of 1.2-2.6 cases per million.<sup>[1]</sup> It is a systemic disease with generalized as well as vascular symptoms. The generalized symptoms include malaise, fever, night sweats, arthralgias, anorexia, and weight loss, which may occur months before vessel involvement is apparent. These symptoms may merge into those related to vascular compromise and organ ischemia. Thickening of the vessel wall is the earliest manifestation of the disease before stenosis and dilatation occur. Color Doppler may be useful in identifying circumferential vessel wall thickening and thereby in evaluating and monitoring the disease in the aorta and its branches. Conventional or digital subtraction angiography is the standard imaging tool for diagnosis and evaluation of vasculitis. But it only demonstrates the appearance of the lumen; it may be normal in the initial stages of the disease with only diffuse mural thickening without any luminal narrowing. In these situations, noninvasive multidetector CT angiography is helpful in demonstrating mural thickening as well as luminal narrowing. Use of contrast material may reveal enhancing inflammatory lesions in the early stage of the disease, prior to the development of stenosis.<sup>[2]</sup> Magnetic resonance (MR) angiography can also demonstrate thickening of the vessel wall, along with better soft tissue differentiation and other signs of inflammation like mural edema and increased mural vascularity.



**Figure 2:** Saggital contrast enhanced computed tomography image showing thickened wall of the descending aorta with narrowing of the caliber (large arrow). Volume-rendered angiographic images shows narrowing of the superior mesenteric artery (small arrow) with stenosis of bilateral renal arteries involving the ostia and proximal segments (double arrows) with post-stenotic dilatation. The arterial branch to the sequestered lung segment from the thoracic aorta is well seen in the central angiographic image

Two types of pulmonary sequestration are recognized, depending on whether the abnormal lung tissue possesses its own pleural covering. In the intralobar pulmonary sequestration, the malformation is incorporated in the normal pulmonary parenchyma of a lobe with the venous drainage via the pulmonary veins. The extralobar pulmonary sequestration consists of pulmonary parenchyma separated from the rest of the lung by its own pleural envelope with its venous drainage via the systemic veins into the right atrium. Pulmonary sequestration remains a rare congenital anomaly comprising  $0.15 \pm 6.4\%$  of all congenital pulmonary malformations<sup>[3]</sup> and  $1.1 \pm 1.8\%$  of all pulmonary resections.<sup>[4]</sup>

Numerous reports have described serious complications arising from pulmonary sequestrations such as fungal infection,<sup>[5]</sup> tuberculosis,<sup>[6]</sup> fatal hemoptysis, massive hemothorax,<sup>[7]</sup> cardiovascular problems,<sup>[8,9]</sup> benign tumors,<sup>[10]</sup> and even malignant degeneration.<sup>[11]</sup> Distinguishing pulmonary sequestration from other pulmonary disorders, such as congenital cystic adenomatoid malformation, pulmonary emphysema, intrapulmonary abscess, bronchiectasis, or malignant tumor is not always possible on chest X-ray as on plain radiograph, the sequestered segment shows heterogeneous opacity with some cystic areas within. Color Doppler is helpful in identification of the feeding artery from the aorta. Three-dimensional (3D) reconstructions with multidetector CT depict the anomalous arterial supply and venous drainage. Angiography is not a part of the routine investigation but is the gold standard in determining arterial supply.

Our patient presented with symptoms of intestinal angina, which included postprandial pain and weight loss, but appetite was intact. She was also incidentally found to have high blood pressure. Both the findings are very well explained with the CT angiography report. The superior mesenteric artery narrowing led to intestinal ischemia, which worsened on food intake due to mismatch of blood supply and demand. The stenosis of renal arteries explained the pathogenesis of hypertension, which was renovascular in origin. Common differential diagnoses like tuberculosis and Takayasu's arteritis were considered early during evaluation. However, her chest X-ray was normal, and tuberculin test was also negative. Moreover, her clinical profile did not meet the

American College of Rheumatology (ACR) criteria for Takayasu's arteritis.<sup>[12]</sup> There was an incidentally detected pulmonary sequestration in our patient. The patient was asymptomatic and had no chest symptoms attributable to sequestration. The patient was started on steroids and antihypertensive agents. After two months on the above-mentioned treatment, her symptoms resolved. Blood pressure was under control.

To the best of our knowledge, this is the first ever case reported to have an asymptomatic intralobar pulmonary sequestration in association with medium and large vessel vasculitis.

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