A 41-year-old woman presented with cough and heart murmur for a half-month. She has hyperthyroidism but is well controlled. The results of laboratory tests, including kidney and liver functions, electrolytes, and general urine, were normal. The erythrocyte sedimentation rate (32 mm/h; normal value, <20 mm/h) and C-reactive protein (35 mg/L; normal range, 0–20 mg/L) were high; hemoglobin (122 g/L; normal value, 110–160 g/L), leukocytosis (white blood cells 8.8×10⁹/L; normal value, 4.8–10.8×10⁹/L), and thrombocytosis (299×10⁹/L; normal value, 100–300×10⁹/L) were normal. Initial blood tests showed that troponin I at 0.44 ng/mL (normal value, 0–0.16 ng/mL), B-type natriuretic peptide at 1630 pg/mL (normal value, 0–400 pg/mL), and creatine kinase at 6.6 ng/mL (normal value, 0–4.0 ng/mL) were elevated. Immunology testing showed that antinuclear antibody lineages were negative.

Chest x-ray film demonstrated clear lung fields but an enlarged heart. ECG showed sinus tachycardia, right axis, and signs of right atrium and right ventricle overload.

Transthoracic echocardiography demonstrated a giant right atrium (56 mm) and right ventricle (57 mm) (Figure 1A and online-only Data Supplement Movie I). The left ventricular size (42 mm) and systolic function (ejection fraction=70%) were normal. The intimal was significantly thickened in the pulmonary artery and branches (Figure 1B). Color Doppler flow imaging showed a turbulence color signal in the left PA and no flow signals within the right PA (Figure 1C, and online-only Data Supplement Movie II); mild tricuspid valve regurgitation with systolic pulmonary artery pressure was estimated at 95 mm Hg (Figure 1D).

Pulmonary artery and venous angiography by 64-slice spiral computer tomography (CT) was performed. Cine imaging demonstrated right PA occlusion and severe left PA stenosis (Figure 2A and 2B).

Exploratory surgery of the pulmonary vascular structure by extracorporeal circulation was performed successfully. Intraoperative findings showed that the right PA wall was significantly thickened and solid leading to luminal obliteration, and the left PA was stenotic, whereas there were no obvious abnormalities in the pulmonary veins. A histopathological examination was performed of the PA wall. Microscopic examination of the tissue biopsies showed granulomatous inflammation in the endometrial tissue of PA, with a large number of plasma cells and fiber necrosis (Figure 3A). Immunohistochemistry staining of the tissue showed polyclonal plasma cell and lymphocyte infiltration (Figure 3B), which was consistent with lymphomatoid granulomatosis. No immunoglobulin deposits were seen.

The histopathologic findings were consistent with isolated pulmonary vasculitis. The patient was treated by the intravenous corticosteroids (20 mg of prednisone every 24 hours for 3 consecutive days) followed by prednisone 20 mg/d and low-dose aspirin. Two weeks later, her clinical symptoms were significantly improved; meanwhile, echocardiographic examination showed that the systolic PA pressure estimated by the velocity of tricuspid regurgitation was significantly decreased (from 95 to 68 mm Hg). The CT Cine imaging demonstrated that the right PA reopened with blood filling and left PA stenosis was improved (Figure 2C). Six weeks later, echocardiography confirmed a normal-size right atrium (36 mm) and right ventricle (36 mm). Estimated systolic PA pressure was 40 mm Hg.

The clinical course of our patient confirmed the benign reactive nature of the plasma cell granuloma after treatment. However, patient was followed after 22 months; she was continuing to take prednisone 5 mg/d, but experienced cough, shortness of breath, and was getting worse. Echocardiographic images displayed severe tricuspid regurgitation and an estimated systolic PA pressure of 145 mm Hg (Figure 4A and 4B), a more enlarged right atrium (61 mm) and right ventricle (64 mm) with dysfunction (online-only Data Supplement Movie III), and right PA occlusion again and more severe left PA stenosis (Figure 4C and online-only Data Supplement Movie IV), which were confirmed by CT scan (Figure 4D). The clinical condition of the patient had significantly declined. Given the lack of response to drug therapy and the progressive clinical deterioration, lung transplantation was suggested, but the patient and her family refused any further intervention and then applied for discharge.

**Discussion**

The vasculitides constitute a group of heterogeneous conditions characterized by blood vessel inflammation and
necrosis, leading to subsequent tissue or organ injury. They are usually systemic diseases affecting multiple territories or organs with overlapping clinical and pathological manifestations. However, there are cases in which inflammation is restricted to a single organ. Isolated pulmonary vasculitis is a rare entity; only a few cases, mostly affecting large pulmonary vessels, have been described.

Although in some of these cases pulmonary vasculitis may be asymptomatic, pulmonary hypertension is a common symptom at presentation that may lead to a false-positive diagnosis of chronic thromboembolic disease. When suspected, pulmonary angiogram, magnetic resonance imaging, or positron emission tomography scan are useful diagnostic tools, typically showing wall thickening, narrowing, or stenosis of the large vessels in patients with isolated large-vessel pulmonary vasculitis.

In this report, we describe a patient with localized pulmonary vasculitis affecting medium-sized vessels that presented as pulmonary arterial hypertension. The clinical conditions of our patient were significantly improved by treatment with intravenous corticosteroids, but did not maintain, which might be because the dosage of prednisone was not enough to control the immunity inflammation. This case is a good example of localized pulmonary vasculitis involving medium-sized vessels. Although conditions such as Behçet syndrome or polyarteritis may share similarities with this case, our patient had no history of oral or genital ulcers, cutaneous lesions, or uveitis that are typical of Behçet syndrome. In addition, apart from the vasculitic pulmonary involvement, there was no evidence of involvement of other organs that is typical of cases of polyarteritis.

The nonspecific nature of the presenting symptoms, the relatively low rate of systemic symptoms and physical signs, and the variety in the pace of disease progression often result in delayed diagnosis of isolated pulmonary vasculitis. Early diagnosis of isolated pulmonary vasculitis is crucial because it allows early aggressive treatment that will predict better response and prevention of irreversible stenotic and fibrotic vascular changes.

Patients with pulmonary artery hypertension caused by pulmonary vasculitis have a poor prognosis and higher rates of death. Echocardiography is a useful noninvasive tool for estimating systolic pulmonary artery pressure using tricuspid regurgitation jet flow. In this case, the occlusion of the right PA and stenosis of the left PA were detected by echocardiography with more careful inspection, and CT images provided more information in detail.

**Disclosures**

None.

**References**

Figure 1. A, Transthoracic echocardiographic 4-chamber view showed marked enlarged right atrium and ventricle. B, Parasternal short-axis view showed significant thickening of the intimal in left and right pulmonary arteries. C, Parasternal short-axis view with color Doppler showed turbulence color signal in the left pulmonary artery (red arrow) and no flow signals within the right pulmonary artery (yellow arrow). D, Continues-wave Doppler obtained from tricuspid valve regurgitation indicated a high velocity, and the estimated systolic pulmonary artery pressure was 95 mmHg. AO indicates aorta; LA, left atrium; LPA, left pulmonary artery; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RPA, right pulmonary artery; and RV, right ventricle.

Figure 2. Computed tomographic pulmonary angiography and volume reconstruction images A, Short-axis view showed the occluded right main pulmonary artery (yellow arrow) and left main pulmonary artery stenosis (red arrow). B, CT reconstruction image showed left pulmonary artery stenosis (red arrow) and the occluded right main pulmonary artery (yellow arrow). C, Marked improvement of LPA stenosis (red arrow) and perfusion recovery of RPA (yellow arrow) after 2 weeks of treatment. AO indicates aortic; CT, computed tomography; LPA, left pulmonary artery; LPV, left pulmonary vein; PA, pulmonary artery; and RPA, right pulmonary artery.
Figure 3. A, Histological examination of the pulmonary artery wall obtained from pulmonary biopsies showed granulomatous inflammation in the endometrial tissue of the pulmonary artery, with a large number of plasma cells and fiber necrosis. B, Immunohistochemistry staining of the tissue showed polyclonal plasma cell and lymphocyte infiltration.

Figure 4. A, Echocardiography confirmed severe tricuspid insufficiency. B, The estimated systolic pulmonary artery pressure was 145 mm Hg. C, The echocardiographic image showed that the RPA was occluded and that the LPA was severely stenotic. D, CT image showed that the right pulmonary artery was almost occluded with a very fine blood flow (yellow arrow) and that left pulmonary artery had stenosis (red arrow). CT indicates computed tomography; LA, left atrium; LPA, left pulmonary artery; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RPA, right pulmonary artery; and RV, right ventricle.
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Movie Legend

**Movie 1.** Transthoracic echocardiogram showing marked enlarged right atrium and right ventricle. Best viewed with Windows Media Player.

**Movie 2.** Color Doppler flow imaging showed turbulence color signal in the left PA and no flow signals within the right PA. Best viewed with Windows Media Player.

**Movie 3.** Echocardiography showed the severe enlarged RA and RV with dysfunction. Best viewed with Windows Media Player.

**Movie 4.** Echocardiographic image displayed right pulmonary artery could not been seen and left PA was severe stenosis. Color Doppler flow imaging showed turbulence color signal in the left PA. Best viewed with Windows Media Player.