Case Report

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Isolated pulmonary artery vasculitis

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Abstract:

We present a unique case of a 26-year-old woman, which first evaluated as sarcoma or thromboembolic events with their clinical and radiological similarities of the pulmonary artery but diagnosed as isolated pulmonary artery vasculitis after endarterectomy with the help of histopathological examinations. After the endarterectomy procedure, the other causes that may lead to vasculitis had been excluded. The patient had the immunosuppressive therapy after the procedure, and her control computerized tomography revealed that the lumen of the pulmonary artery was wide open. Isolated pulmonary artery vasculitis must be considered as a differential diagnosis when a filling defect of the pulmonary artery had been detected without other systemic findings.

Keywords:

Large-vessel vasculitis, pulmonary artery, single-organ vasculitis

Introduction

The vasculitides are a set of related disorders characterized by blood vessel inflammation and necrosis, leading to tissue or end-organ dysfunction. Classification of these heterogeneous group of diseases can be based on the size (large-sized, medium-sized, or small-sized) of affected vessels and being systemic or isolated.[1] Generally, systemic forms that affect multiple organs are more prevalent. It is said that for isolated forms, "single-organ vasculitis" is a better terminology to describe the situation.[2] Single-organ vasculitides that affect the components of the central and peripheral nervous system, kidney, breast, aorta, and gastrointestinal and urinary systems have been reported.[1] While the aorta is the most common site for large-vessel vasculitis, isolated vasculitis of the pulmonary artery is very rare. In these cases, definitive diagnosis is made on surgical biopsy or autopsies. In literature, reported cases of isolated pulmonary artery vasculitis are infrequent.[3-5]

In this article, we present a unique case of a 26-year-old woman, which first evaluated as sarcoma of the pulmonary artery but later diagnosed as isolated pulmonary artery vasculitis with the help of histopathological examination.

Case Report

A 26-year-old female admitted to our outpatient clinic with complaints of left-sided cramping-like chest pain, exertional dyspnea, and cough with 6 months of duration and one episode of hemoptysis. She had no fever and weight loss. She had no systemic disease and was a nonsmoker. Her physical examination was normal. Laboratory investigations were C-reactive protein 27.5 mg/L (0–5 mg/L), erythrocyte sedimentation rate 35 mm/h, white blood cell count 8.2 × 10^9/L (4.0–

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10.0 × 10^9/L), hemoglobin 11.1 g/dL (12.0–17.0), platelets 557/mm^3 (150–440), D-dimer 2.22 mg/L (0—0.5), pro-B-type natriuretic peptide 38.16 ng/L (0–125). No specific abnormality has been found in other biochemical investigations, including liver and kidney function tests. In posteroanterior chest radiography, there was left hemidiaphragmatic elevation and left lung had volume loss [Figure 1]. According to the Wells' Scoring system, she had a low clinical probability for pulmonary embolism, but she had a high D-dimer level, so we performed a computed tomography angiography (CTA) scan. The CTA revealed left-sided minimal pleural effusion and thickening and a mass lesion (3 cm of its widest area) that almost totally obstructs the lumen of the left pulmonary artery and follows its branches [Figure 2]. Cardiac evaluation and transthoracic echocardiography were normal. Due to suspicion for malignancy, F-fluorodeoxyglucose positron emission tomography-CT (FDG-PET-CT) was performed [Figure 3]. The lesion had 11 SUV_{max} uptake of FDG. There was no other pathological uptake. The patient operated due to the suspicion for sarcoma. During the surgical operation, surgeons approached the mass lesion started from the proximal side of the left pulmonary artery and transmural invasion was seen. Endarterectomy operation was performed to the upper and lower branches of the pulmonary artery, respectively [Figure 4]. On pathological examination, the vessel wall was almost totally infiltrated with mononuclear cells (predominantly with lymphocytes), and accumulated infiltrations of giant cells and histiocytes were seen and interpreted as vasculitis [Figure 5]. For the patient diagnosed with vasculitis, additional blood tests were performed and the results were as follows: proteinase-3 anti-neutrophil cytoplasmic antibodies (ANCAs) 2.23 RU/mL (0–19), myeloperoxidase-ANCA 3.75 RU/mL (0–19), ferritin 61.20 (5.8–274), anti-beta-2 glikoprotein IgM 19.6 RU/mL (0–19), anti-beta-2 glikoprotein IgG RU/mL 2.81 (0–19), and lupus anticoagulant 35.3 sn (31.4–43.4). Hepatitis B and C and HIV serology were negative. Ear-nose-throat, nephrology, and ophthalmology examinations for other systemic vasculitides revealed no other vasculitic involvement, so the case accepted as isolated pulmonary artery vasculitis. Methylprednisolone (32 mg) and

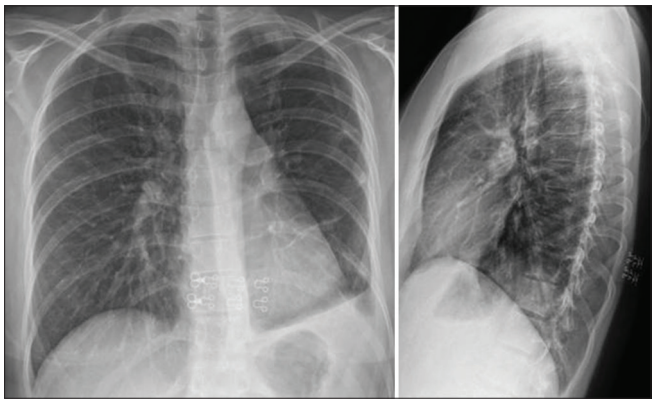

Figure 1: Chest radiographs at the first admission to the clinic

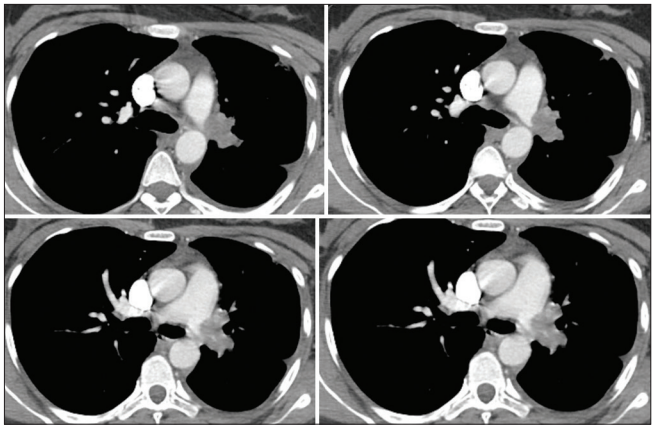

Figure 2: Thorax computed tomography angiography: Mass lesion (3 cm of its widest area) that almost totally obstructs the lumen of the left pulmonary artery and follows its branches

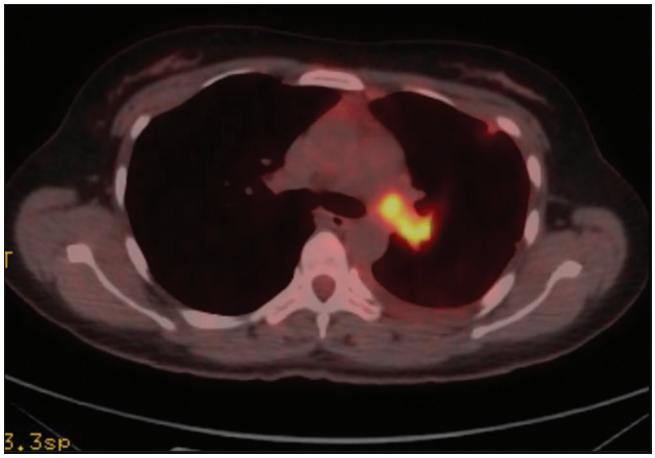

Figure 3: Fluorodeoxyglucose positron emission tomography: The mass lesion of the pulmonary artery with 11 SUV_{max} uptake

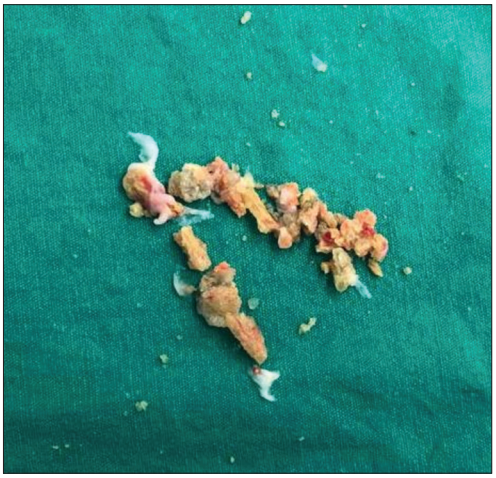

Figure 4: Endarterectomy of the pulmonary artery and arteries of upper and lower lobes

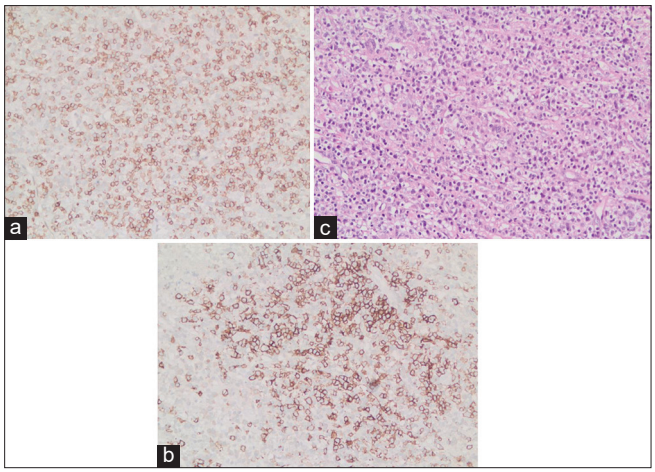

Figure 5: (a) Some of the lymphocytes positive for CD3 (IHC, ×20), (b) some of the lymphocytes positive for CD20 (IHC, ×20), (c) mononuclear inflammatory cells infiltrating vessel wall, consisting mainly lymphocytes, scattered plasma cells and histiocytes (H and E stain, ×20)

azathioprine were started as treatment. The patient is still under treatment; however, on control computed chest tomography, lumen of pulmonary arteries appeared totally open [Figure 6].

Discussion

In this case, the proximal side of the left pulmonary artery was almost totally obstructed with a solid mass lesion. Due to the suspicion for malignancy on PET/CT, endarterectomy was performed to the patient for definitive diagnosis. With the help of pathological and clinical examinations, the case evaluated as isolated pulmonary artery vasculitis.

The most prevalent cause of pulmonary artery obstruction is thromboembolism.[6] Other causes can be listed as sarcoma, vasculitis, dirofilariasis, and congenital pulmonary atresia.[6,7] The incidence of isolated pulmonary vasculitis is not known but thought to be infrequent. In literature, very few cases have been reported. Clinical and radiological features of pulmonary artery vasculitis can be similar to malignancy and chronic thromboembolism.[3,4] In these cases, the obstructive solid lesion in the radiological examination may unexpectedly diagnosed with vasculitis.[2] Kitajima *et al*. reported a case of a 42-year-old patient who underwent lobectomy due to lack of enough evidence to exclude sarcoma.[7] In another case, mediastinoscopy and open lung biopsy have been performed with suspicion for malignancy.[4] In some cases, pulmonary hypertension is misinterpreted as chronic thromboembolism, and treatment and follow-up have done accordingly. Hagan *et al*. reported two cases who underwent endarterectomy due to not responding to the chronic thromboembolism treatment and diagnosed as vasculitis with the histopathological examinations.[3] In

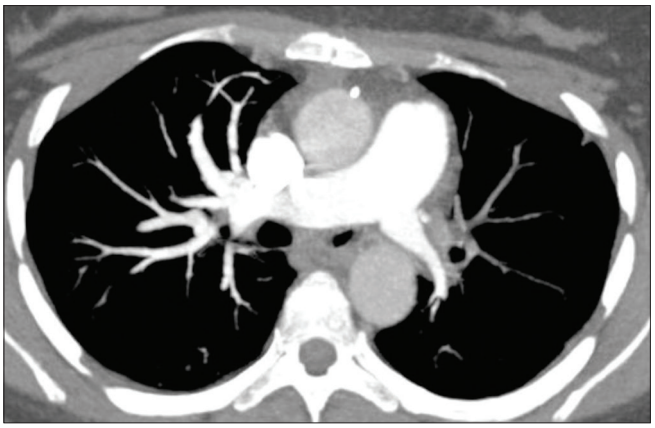

Figure 6: The lumen of the pulmonary artery is totally open on control chest computed tomography

our case, due to the high FDG uptake in PET-CT imaging, the patient underwent surgical biopsy. During the operation, the gross features of the lesion were consistent with sarcoma. After the endarterectomy procedure, the histopathological examination revealed vasculitis.

Other vasculitic diseases that involve pulmonary artery include systemic vasculitic diseases such as granulomatosis with polyangiitis (formerly known as Wegener granulomatosis), eosinophilic granulomatosis with polyangiitis (formerly known as Churg-Strauss syndrome), Behçet's disease, and polyarteritis nodosa. In our case, there was a large-vessel vasculitis. ANCA was negative. There were no aphthous ulcers on the genitalia, no skin lesions, and no other systemic signs and symptoms. These features led us to exclude the systemic vasculitic diseases listed above. Large-vessel involvement can be seen in Takayasu arteritis and giant cell arteritis, but pathological examination of our case revealed no granulomas which are usual pathological features of these vasculitic diseases.

As diagnostical radiological imaging techniques, chest tomography with contrast material, magnetic resonance imaging, and PET can be used to visualize the thickening, narrowing, and stenosis of the vessel wall.[8] In literature, in addition to the cases with medium metabolic activity on PET-CT scan, there are also cases with no metabolic activity.[4,7] In our case, the lesion had high metabolic activity on PET/CT.

Early diagnosis is important for better response to the treatment and the prevention of irreversible and fibrotic vascular changes. Prognosis may be poor and mortality may be high when pulmonary hypertension and aneurism emerged. Moghaddam *et al*. reported a case of a 74-year-old with isolated pulmonary artery vasculitis whose pulmonary artery aneurysm progressed so had to be operated.[9] In other reported cases of two young people, lung

transplantation is considered due to advanced pulmonary hypertension (pulmonary artery pressure approximately 95–100 mmHg) and one of them underwent bilateral lung transplantation.[5]

The first-line treatment is surgical resection. The first choice in medical treatment is corticosteroids, then immunosuppressive agents such as cyclophospomide, azathioprine, mycophenolate mofetil and infliximab can also be used. For the cases with pulmonary hypertension, vasodilators (sildenafil, bosentan) and rituximab need to be added to the treatment.[3,4] Although the response to the treatment in the early course of the disease was excellent, the vessels of other parts of the body can also be affected during the follow-up. These can be interpreted as recurrence and may be progressive.[3,5] There are cases of pulmonary vasculitis which underwent a lung transplant.[4] In our case, after endarterectomy, methylprednisolone and azathioprine treatment has been started, and on control CT imaging, the lumens of the pulmonary arteries appeared all open at the 3rd month of follow-up.

In conclusion, patients with filling defects in pulmonary vasculature and isolated pulmonary artery vasculitis without systemic features should be kept in mind. In these cases, for definitive diagnosis, surgical resection should be considered due to suspicion for malignancy.

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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