Pulmonary Artery Compression and Invasion by a Ruptured Giant Thoracic Aortic Aneurysm: A Rare Presentation

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INTRODUCTION

Giant thoracic aortic aneurysm (TAA) is a life-threatening condition that carries a high risk for rupture and may cause symptoms and complications by compressing adjacent structures. Although rare, extrinsic compression of the pulmonary artery (PA) may mimic pulmonary thromboembolism (PTE) and is most commonly associated with ascending aortic aneurysm. There are only few reports of rupture of TAA into the PA, and they are usually of inflammatory etiology.

Here we present a case of severe pulmonary hypertension (PH) and right ventricular failure (RVF) secondary to stenosis of the main PA caused by an intraluminal thrombus, resulting from a ruptured giant TAA, extending to the right PA and complete occlusion of the left PA. To our knowledge this is the first report of invasion of the PA by a ruptured giant TAA causing occlusion of the left PA and extrinsic compression with severe stenosis of the main PA, without the development of aortopulmonary fistula.

CASE PRESENTATION

A 32-year-old male patient with a 2-month history of abdominal pain, fever, and progressive dyspnea was initially admitted to a general hospital with main diagnostic hypothesis of abdominal sepsis. After diagnostic workup, no signs of infection were found, and a giant TAA was diagnosed on thoracic computed tomography, leading to transfer to our tertiary care hospital for cardiac surgical treatment. The patient was previously healthy, with no family history of cardiovascular or immunologic diseases. Physical examination at admission revealed regular pulse, clear lungs, and hypophonetic heart sounds, without appreciable heart murmurs. There were signs of marked systemic venous congestion, with dilated vena cava and jugular distension. Electrocardiography showed sinus rhythm and incomplete right bundle branch block without primary repolarization abnormalities. Transthoracic echocardiography was performed upon admission, revealing signs of severe PH, RVF, a large pericardial effusion (Figure 1, Video 1), and dilated inferior vena cava. A mobile mass was seen invading the main PA (Figure 2, Video 2) in continuity with a large TAA (10.3 cm in its major diameter) involving the ascending aorta and aortic arch (Figure 3, Videos 3 and 4). There was a severe narrowing caused by the intraluminal mass inside the PA (Figure 4A, Video 5), with a peak systolic gradient of 98 mm Hg (Figure 4B), and no flow was detected in the left PA, suggesting complete occlusion. The findings were very suggestive of contained wall rupture of the aortic aneurysm into the PA, extending to the right PA. On three-dimensional echocardiography, the continuity between the intraluminal thrombus and the aortic aneurysm was very nicely depicted (Figure 5A, Video 6), and the severe narrowing of the main PA by the large mobile thrombus was also seen in details (Figure 5B, Videos 7 and 8). Computed tomographic angiography confirmed the echocardiographic findings, showing rupture of the aortic aneurysm into the PA (Figures 6 and 7) and, on three-dimensional rendered reconstructed images, the morphology of the thoracic aorta, narrowing of the main PA, and occlusion of the left PA (Figure 8). Serologic tests for syphilis and hemocultures in the preoperative evaluation were all negative. The patient underwent surgery, including aneurysm resection, pulmonary thromboendarterectomy (with main PA and right PA reconstruction with bovine pericardial patch), partial replacement of the aorta with supracoronary aortic graft (Dacron 26 mm), and reimplantation of the brachiocephalic trunk and left carotid artery, with the distal anastomosis of the graft directed to the descending thoracic aorta (intended to be followed by a staged thoracic endovascular aortic repair; Figure 9).

In the immediate postoperative period the patient developed fever, leukocytosis, and high levels of C-reactive protein and lactate. Broad-spectrum empiric antibiotics were initiated, and *Serratia marcescens* KPC was later isolated from hemocultures. Despite antibiotics and intensive support with vasoactive amines and volemic reposition, the patient developed septic shock (tachycardia and hypotension with an inflammatory profile on Swan-Ganz catheterization) and renal, respiratory, and hepatic insufficiency, culminating in death due to multiple organ failure. Histopathologic examination of the anatomic specimen showed intima and media necrosis of the aortic wall, fibrosis, and inflammatory infiltrate with damage to the elastic component of the media, an aspect of necrotizing aortitis (NA). Tissue cultures were all negative, and no other source of sepsis was found (Figure 10).

DISCUSSION

Most TAAs are caused by degenerative disease, resulting in dilatation of the aorta. The incidence of TAA is estimated to be increasing, and there are about 10.4 cases per 100,000 person-years.[1] Giant TAAs are defined as dilatations >10 cm and are associated with a greater risk for rupture and compression of mediastinal structures.[2] In the majority of cases, TAAs have an asymptomatic course and are found incidentally

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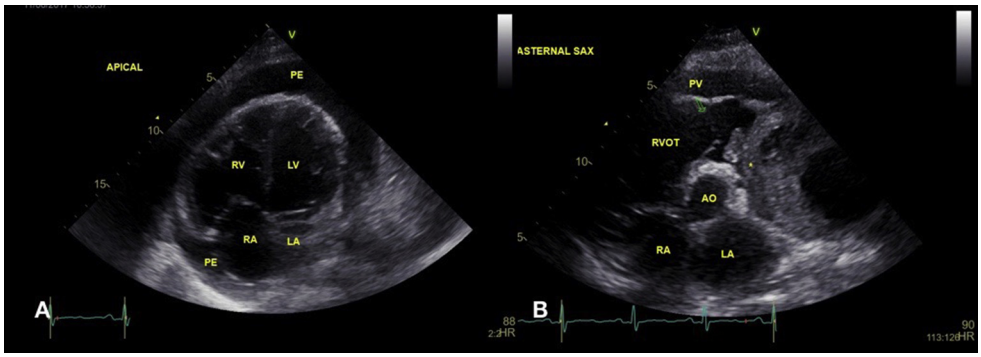

Figure 1 **(A)** Transthoracic echocardiography. Apical four-chamber view. Right ventricular and right atrial dilatation, right ventricular pressure overload and dysfunction, and severe pericardial effusion (PE). **(B)** From the parasternal short-axis (SAX) view, a large mobile thrombus is seen inside the main PA, causing severe stenosis. *AO*, Aorta; *LA*, left atrium; *LV*, left ventricle; *PV*, pulmonary valve; *RA*, right atrium; *RV*, right ventricle; *RVOT*, right ventricular outflow tract.

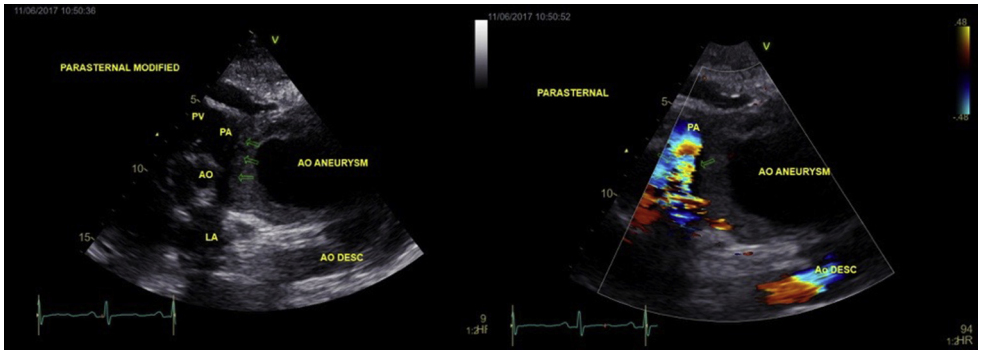

Figure 2 Transthoracic echocardiogram. Modified (higher) parasternal short-axis view showing the large mass invading the main PA (thrombus) in continuity with the large TAA (AO ANEURYSM). The mass causes severe stenosis of the PA (*arrows*). *AO*, Aorta; *AO DESC*, descending thoracic aorta; *LA*, left atrium; *PV*, pulmonary valve.

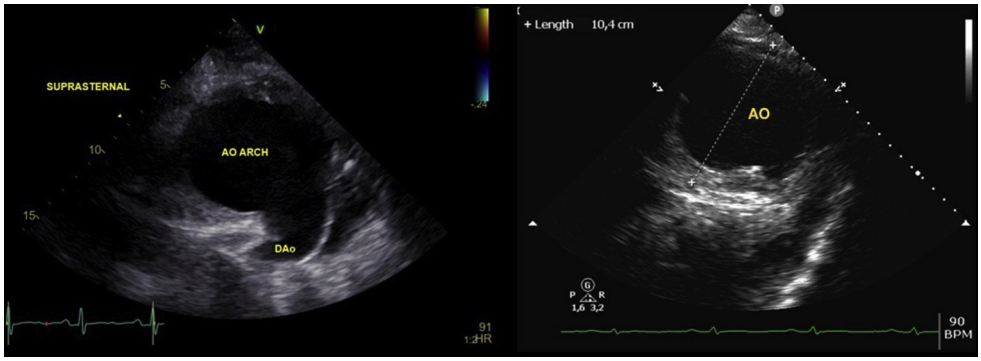

Figure 3 Transthoracic echocardiography. Suprasternal view depicting the large TAA involving the distal segment of the ascending aorta (AO) and the aortic arch (AO arch). The aneurysm measures 10.4 cm in its major diameter. *DAo*, Descending aorta.

on routine radiography. They may cause symptoms by compression of adjacent structures, including hoarseness (left recurrent laryngeal nerve compression), stridor (bronchial compression), dyspnea (lung compression), dysphagia (esophageal compression), and plethora (superior vena cava compression). In some cases aortic valve regurgitation due to aortic root or ascending aortic dilatation is seen. The great majority of TAAs are in the ascending portion, and because of the intimate anatomic relationship to the PAs, extrinsic compression by mass effect can be seen[3,4] and in rare cases even rupture into the PA. It is well described that these two vessels share a common "sheath" of mediastinal connective tissue.[5] Any rupture of the aortic adventitia may cause hemorrhagic infiltration along this tissue, which may erode the wall of the PA, causing rupture, aortopulmonary fistulas,[6] and in some cases occlusion of the PA.[7]

Compression of the PA by TAA is rare and was reported in the literature more frequently in the prewar period, when syphilitic aortitis was more prevalent. This extrinsic compression may cause PH and RVF, clinically mimicking PTE. Although antibiotic use dramatically decreased the incidence of syphilis, some cases of extrinsic compression caused by syphilitic TAA are described in the literature, so this etiology must be considered a potential cause, especially in developing countries.[8] Other aortic infectious diseases, aortic dissection, aortic

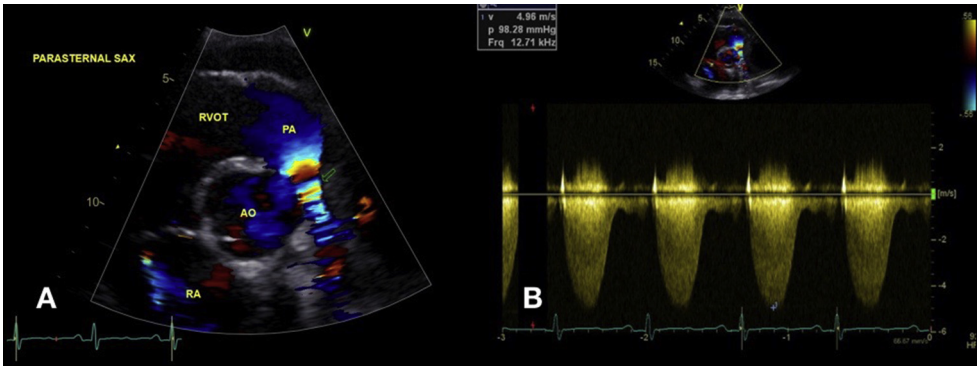

Figure 4 **(A)** Transthoracic echocardiography. Parasternal short-axis view with color Doppler, showing severe stenosis of the main PA. **(B)** On continuous Doppler, a peak systolic gradient of 98 mm Hg is seen, confirming severe PH. *AO*, Aorta; *RA*, right atrium; *RVOT*, right ventricular outflow tract.

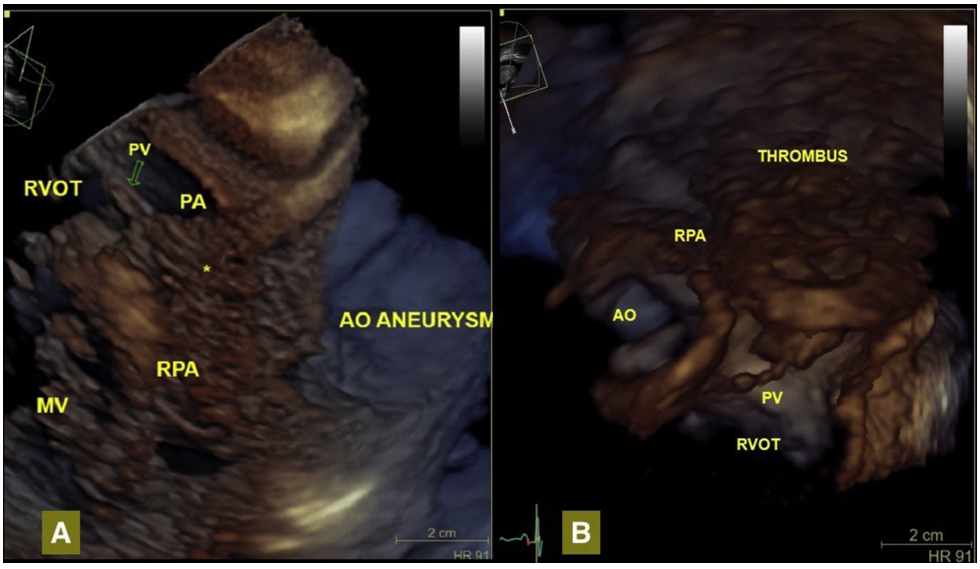

Figure 5 Transthoracic three-dimensional echocardiogram. Data sets acquired in full-volume multibeat acquisition. **(A)** An oblique view showing the continuity between the intraluminal pulmonary mass and the TAA. **(B)** The large mobile thrombus inside the main PA in the supravalvar region, causing severe stenosis. *AO*, Aorta; *AO ANEURYSM*, TAA; *MV*, mitral valve; *PV*, pulmonary valve; *RPA*, right PA; *RVOT*, right ventricular outflow tract.

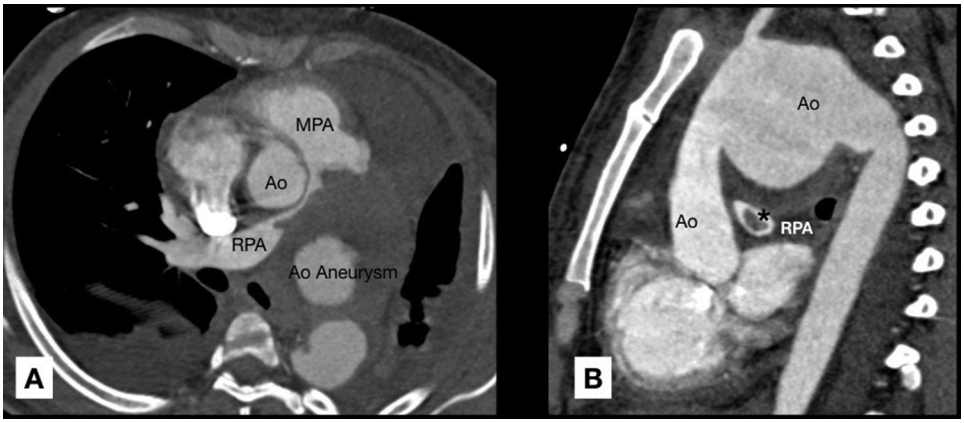

Figure 6 Computed tomographic thoracic angiogram. **(A)** Short-axis view showing the large TAA (Ao Aneurysm), with signs of rupture into the main PA (MPA) and extending to the right PA (RPA), causing severe stenosis. The left PA is occluded. **(B)** Sagittal view showing the entire extension of the thoracic aorta (Ao), with aneurysm involving the distal ascending portion and arch and the thrombus extending toward the PA, with invasion of the RPA well defined in a cross-section of the vessel (*asterisk*).

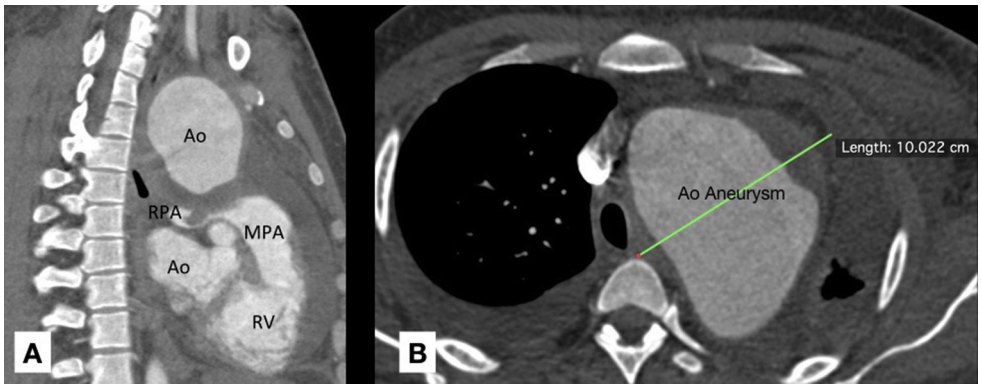

Figure 7 Computed tomographic thoracic angiogram. **(A)** Sagittal view showing the invasion of the main PA (MPA) and right PA (RPA) by the thrombus, caused by a ruptured TAA. **(B)** Short-axis view with measurement of the large TAA of 10.02 cm. *Ao*, Aorta; *RV*, right ventricle.

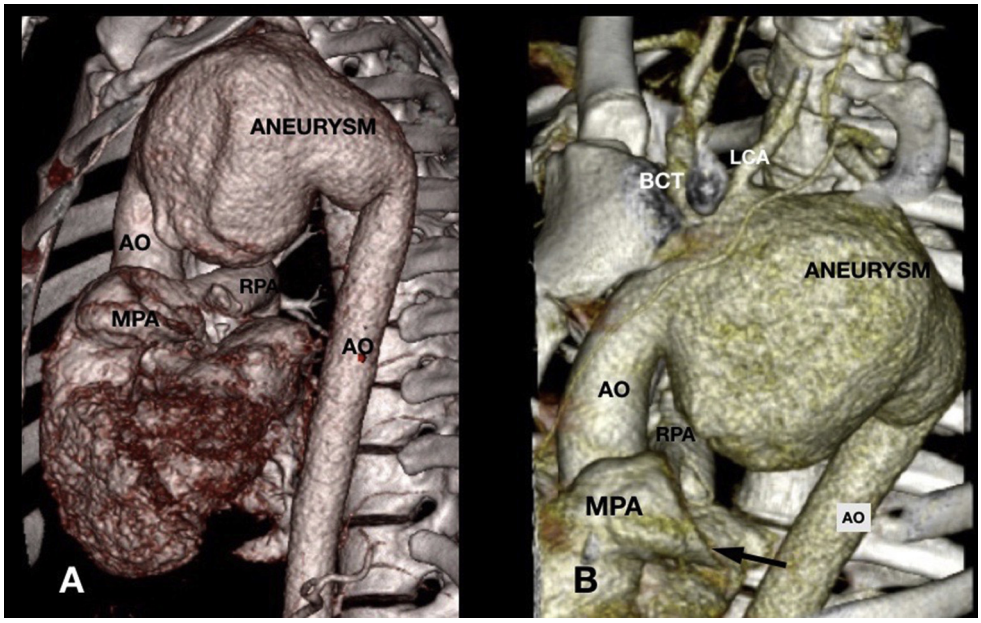

Figure 8 Surface-rendered computed tomographic thoracic angiogram. **(A)** Left lateral view showing TAA and its relation to adjacent structures (note that the thrombus is not visible between the aorta [AO] and the main PA [MPA] because the vascular images are reconstructed as a "luminogram," detecting only contrasted areas inside the vessels and cardiac chambers). **(B)** Oblique view, between anteroposterior and left lateral view, showing the origin of the brachiocephalic trunk (BCT), left carotid artery (LCA), and the large TAA involving the distal ascending aorta and the arch. In both images, severe narrowing of the MPA and right PA (RPA; *black arrow*), with total occlusion of the left PA, is seen.

hematoma, and large noninfectious TAAs are also potential causes.[9,10] Of the cases of aortic aneurysm rupture, the vast majority involve the ascending aorta,[11] and in fact only 3% arise from the aortic arch.[6]

Aortitis is characterized as inflammation of the aortic adventitia and media layers. Rarely it may be caused by infectious diseases (syphilis, *Salmonella*, *Staphylococcus*, and *Mycobacterium*) and is most frequently caused by noninfectious inflammatory and autoimmune diseases. The most common causes of aortitis are Takayasu arteritis and giant-cell arteritis, but it may also occur in association with other rheumatologic disorders or as an isolated idiopathic form.[12] Isolated aortitis more frequently has an asymptomatic course, is clinically underdiagnosed, and may be an incidental finding at the time of histopathologic review of resected aortic aneurysm specimens.[13] In a retrospective study, Liang *et al.*[14] reviewed patients with histologic evidence of active noninfectious aortitis who underwent ascending aortic aneurysm resection at the Mayo Clinic, and they found that the majority (81%) were isolated variant.

Burke *et al.*[15] proposed a histologic classification of noninfectious aortitis, dividing it into two categories: NA and non-necrotizing aortitis. They proposed that NA is an autoimmune condition that may be localized (isolated NA) or may be part of a systemic autoimmune process. NA has a histopathologic appearance of zonal medial laminar necrosis, degeneration, and destruction of the elastic lamina.

Our patient had no report of previous diseases, and all tests for other immunologic and autoimmune diseases were negative, pointing to a case of isolated NA.

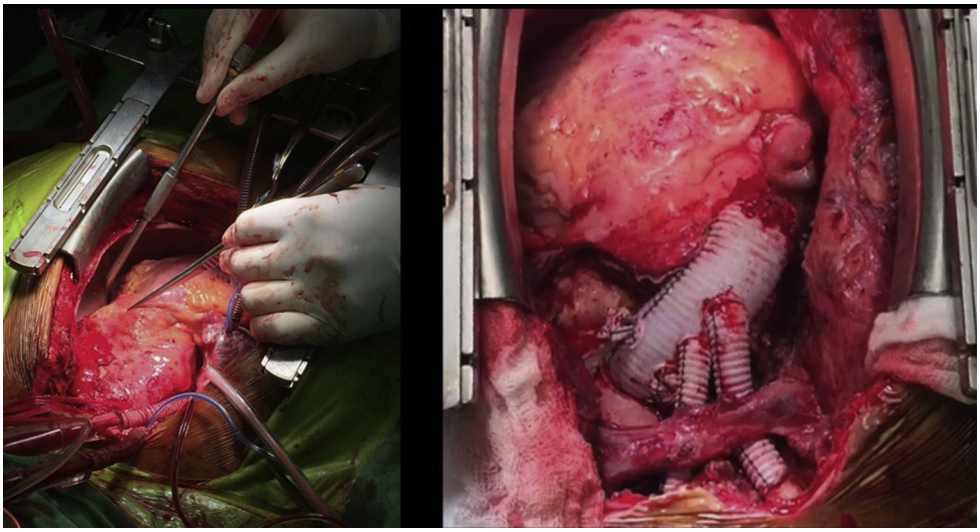

Figure 9 Intraoperative images (surgical view). (*Left*) On surgical inspection, a large TAA was found, involving the ascending aorta and aortic arch. (*Right*) The final operative results, with supracoronary aortic graft implant (Dacron 26 mm) and reimplantation of the brachiocephalic trunk and left carotid artery, with the distal anastomosis of the graft directed to the descending thoracic aorta.

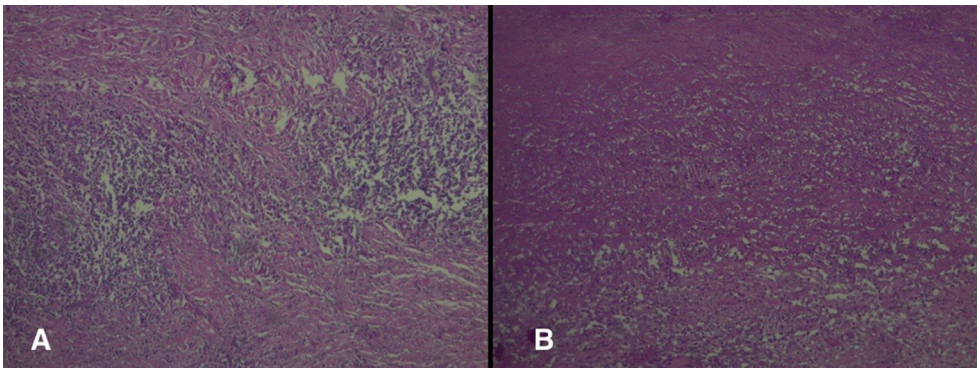

Figure 10 Pathologic specimen of the aortic wall. Hematoxylin and Eosin stain, with magnification of 10×. **(A)** An inflammatory infiltrate, with predominance of mononuclear cells. **(B)** Necrosis of the media, with cellular debris and neutrophils.

CONCLUSION

Although pulmonary embolism is the first diagnosis that comes to mind in patients with severe PH and RVF, mass effect of a giant TAA extrinsically compressing the PA with rupture into the PA must also be considered for the differential diagnosis.

The findings on transthoracic echocardiography, further characterized by three-dimensional echocardiographic images and reconstructed thoracic computed tomographic angiography, were all confirmed on surgical inspection. Histopathologic examination established the etiologic diagnosis of necrotizing aortitis.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.02.005.

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