Intrapericardial Aortic Pseudoaneurysm: A Rare Masquerade of Constrictive Pericarditis

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INTRODUCTION

Aortic pseudoaneurysm or "false" aneurysm is defined as a dilatation of the aorta due to disruption of all wall layers, of which the communicating hematoma is only contained by the periaortic connective tissue.[1] The exact incidence of aortic pseudoaneurysm is uncertain, but its occurrence is almost always anteceded by noxious triggers such as vascular surgery, blunt trauma, infection, or iatrogenicities. Spontaneous occurrence is rare but not unprecedented.[2] We report a case of pseudoaneurysm arising from the aortic root that was entirely confined within the pericardium. The tamponade effect exerted within the pericardial sac produced a unique hemodynamic alteration mimicking that of constrictive pericarditis.

CASE PRESENTATION

A 43-year-old woman presented with nonspecific chest discomfort and progressive worsening of exercise tolerance (New York Heart Association class II) for the past 6 months. She was treated for smear-positive pulmonary tuberculosis 3 years prior without residual complications. She was otherwise healthy and not hypertensive. No preceding chest trauma was reported. Family history was negative for genetic disorders of connective tissue such as Marfan's syndrome. Physical examination was largely unremarkable. The patient was pink and normotensive with equal peripheral pulses. No pulsus paradoxus or Kussmaul's sign were present. A 6-minute walk test recorded 425 m. Blood investigations were all within normal limits. Screening for human immunodeficiency virus and pulmonary tuberculosis were negative. Electrocardiogram showed sinus rhythm and normal axis and complexes.

Chest radiograph (Figure 1) showed cardiomegaly with abnormal right cardiac contour suggestive of enlarged right cardiac chambers or perihilar mass. Otherwise, no evidence of pericardial calcification was seen. Lung fields were clear.

Transthoracic echocardiography showed normal cardiac chambers. Left ventricular wall thickness was within normal limits with no evidence of myocardial infiltration. Left ventricular ejection fraction was 62% using the biplane method. Visualization of the aortic root was suboptimal (Figure 2, Videos 1 and 2). No significant valvular dysfunction was detected. Interventricular septum appeared hyperkinetic, hinting of positive "septal bounce" (Figure 3 and Videos 1, 3-5). Spectral Doppler showed increased respiratory variation of mitral and tricuspid flow velocity at 36.6% (Figure 4) and 52.4%, respectively (normal, mitral <25%; tricuspid, <40%). Tissue Doppler imaging showed increased left ventricular medial annular velocity (e′: 13 cm/sec) but negative for annulus paradoxus. Otherwise, inferior vena cava diameter was normal at 20 mm and collapsing more than 50%. No thickened pericardium or effusion was present.

A preliminary diagnosis of constrictive pericarditis was made in light of preceding tuberculosis infection. Cardiac catheterization was pursued for confirmation of diagnosis. Invasive pressure tracings showed equalization of diastolic filling pressures in the left and right ventricles consistent with constrictive pericarditis (Figure 5). Accentuated early rapid ventricular filling gave rise to deep and rapid filling wave descent, hence the hallmark "dip-and-plateau" or "square root" sign. These findings collectively indicated ventricular diastolic impairment likely due to reduced pericardial compliance.

The investigation took an abrupt turn when we attempted diagnostic coronary angiography. Unintentional diagnostic catheter manipulation cannulated the neck of a large aneurysmal sac arising from the right lateral aspect of the aortic root. Forceful contrast injection was avoided due to concerns of thrombus embolization and rupture. Fluoroscopy of contrast follow-through from the superior vena cava outlined the silhouette of the aneurysmal mass, which was entirely confined within the pericardium (Figure 6). The adjacent right atrium and ventricle were squashed and displaced caudally. The left coronary artery was normal and unaffected. The right coronary artery was compressed at the ostium and appeared elongated at its proximal segment. The vessel contour was otherwise smooth with thrombolysis in myocardial infarction 3 flow (Figure 7). Hence, a revised diagnosis of intrapericardial ascending aortic pseudoaneurysm was made. The hemodynamic alteration akin to constrictive pericarditis was explained by the tamponade effect exerted by the aneurysmal mass within the confined pericardial space.

Contrast-enhanced computed tomography of the thorax confirmed the presence of a large pseudoaneurysm arising from the aortic root preserving the aortic annulus, measuring 84 mm (anterior-posterior, AP) × 86 mm (transverse, TR) × 72 mm (craniocaudal, CC). The aneurysmal sac was contrast enhanced with filling defects seen on the mural surface suggestive of thrombus. A narrow neck was visualized communicating the pseudoaneurysm hematoma to the aortic lumen (Figures 8 and 9). Three-dimensional volume rendering imaging showed the displaced adjacent structures in relation to the lesion (Figures 10 and 11). The ascending aorta was dented by external compressive force but was otherwise within normal caliber. The arch and descending aorta were unremarkable. No evidence of leak or rupture was reported.

Subsequently, the patient underwent thoracotomy for surgical resection of pseudoaneurysm and aortic root repair. Intraoperatively, the pseudoaneurysm wall was found densely adhered to the adjacent pericardium. The aortic root defect was identified. The heart was compressed and laterally displaced. All other surrounding structures were

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VIDEO HIGHLIGHTS

Video 1: Parasternal long-axis view (single loop): normal left atrial and ventricular sizes. No evidence of obvious pericardial thickening. The aortic root is not well visualized.

Video 2: Apical four-chamber view (single loop): preserved left ventricular systolic function. Right atrium and ventricle appear marginally dilated but within acceptable limits.

Video 3: Parasternal long-axis view (long loop): prominent septal hyperkinesia correlating with respiratory cycles suggestive of 'septal bounce'.

Video 4: Parasternal short axis view (long loop): hyperkinetic interventricular septum.

Video 5: Apical four-chamber view (long loop): bouncing interventricular septum accounting for varying left and right ventricular volumes.

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otherwise grossly normal. Excision of pseudoaneurysm and partial pericardiectomy were done followed by patch repair of the ascending aorta. Histopathological examination confirmed the diagnosis of pseudoaneurysm with complete disruption of vessel wall (Figure 12). Microscopically, only nonspecific inflammatory changes were reported with areas of fibrinoid necrosis. The search for granuloma or malignancy was negative. Postoperatively, patient recovered well and was discharged home uneventfully. Follow-up transthoracic echocardiography indicated normalization of hemodynamic measurements.

DISCUSSION

Thoracic aortic aneurysm is increasingly recognized as a silent killer as only 5% of the affected individuals are clinically symptomatic until late complications strike.[3] In contrast, aortic pseudoaneurysm is unique as the pulsatile hematoma escapes the vascular lumen through a defect on the vessel wall but contained by the surrounding perivascular adventitia or matrix. This in turn may lead to catastrophic vessel rupture with a near 100% fatality rate.[4] Early detection and timely intervention are the keys to optimal outcome. Transthoracic echocardiography is the recommended first-line investigation for all thoracic aortic diseases, which should be made a routine practice in clinical workflow.[5] However, many patient and technical limitations make optimal assessment impossible, especially visualization of deep structures within the rib cage. In the event of uncertainty, a multimodality imaging approach should be advocated.

Our case illustrated how a considerably sized intrathoracic mass evaded surface echocardiographic detection. Its strategic retrosternal position behind the "blind spot" of transthoracic echocardiography made direct visualization challenging and thus markedly reduced the sensitivity of identification. However, distinctive hemodynamic shifts were well evidenced as the mass effect progressively compromised the pericardial compliance, impeding ventricular diastolic refilling. Beyond the hinge point where ventricles failed to accommodate further blood volume, a phenomenon mimicking constrictive pericarditis or cardiac tamponade emerged. Similar observations were also

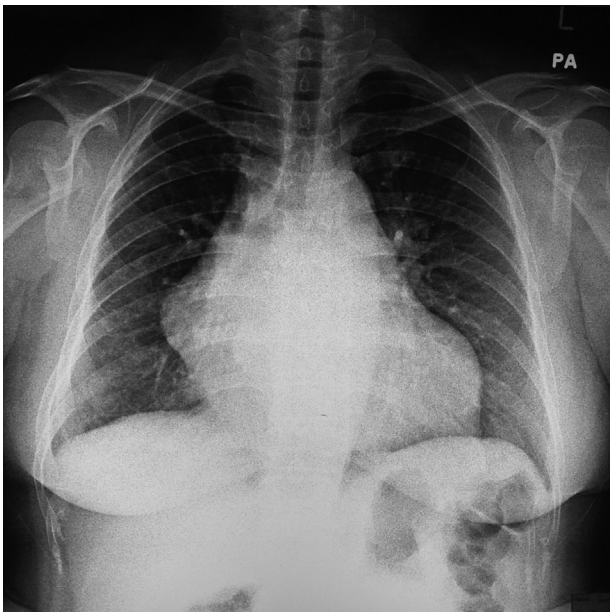

Figure 1 Chest radiograph (posterior-anterior erect). Cardiomegaly with protruding right cardiac contour is suggestive of enlarged right cardiac chamber or mediastinal mass. Lung fields are clear.

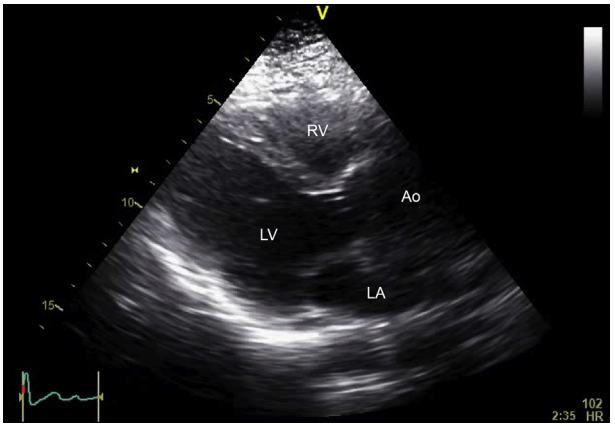

Figure 2 Transthoracic echocardiography (parasternal long-axis view). Aortic root is not well visualized. Cardiac chambers are normal with preserved left ventricular systolic function. No obvious pericardial thickening or effusion is seen. *Ao*, aortic root; *LA*, Left atrium; *LV*, left ventricle; *RV*, right ventricle.

reported in other situations such as infiltrative tumors and organized hematoma without the presence of pericardial effusion.[6,7]

Cardiac catheterization remains the gold standard diagnostic tool for constrictive pericarditis especially when noninvasive tests are inconclusive.[8] Equalization of left- and right-sided diastolic pressures and accentuated early rapid ventricular filling are the hallmark hemodynamic characteristics of constrictive pericarditis. However, it is important to highlight that such observations merely represent the converging consequence of hemodynamic interplay between various factors but not an etiological diagnosis by themselves. Complete resolution can only be achieved if the causal mechanism is identified and addressed conclusively.

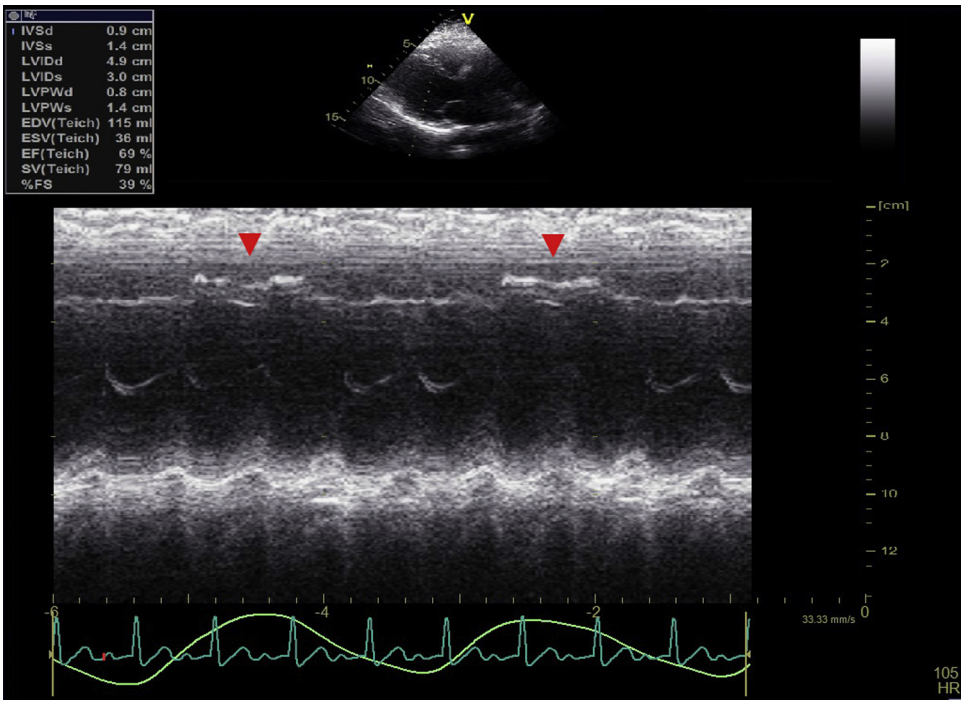

Figure 3 Transthoracic echocardiography (M-mode). Increased ventricular interdependency leads to hyperkinetic septal movement or ''septal bounce'' (*red arrows*) varying throughout respiratory cycles.

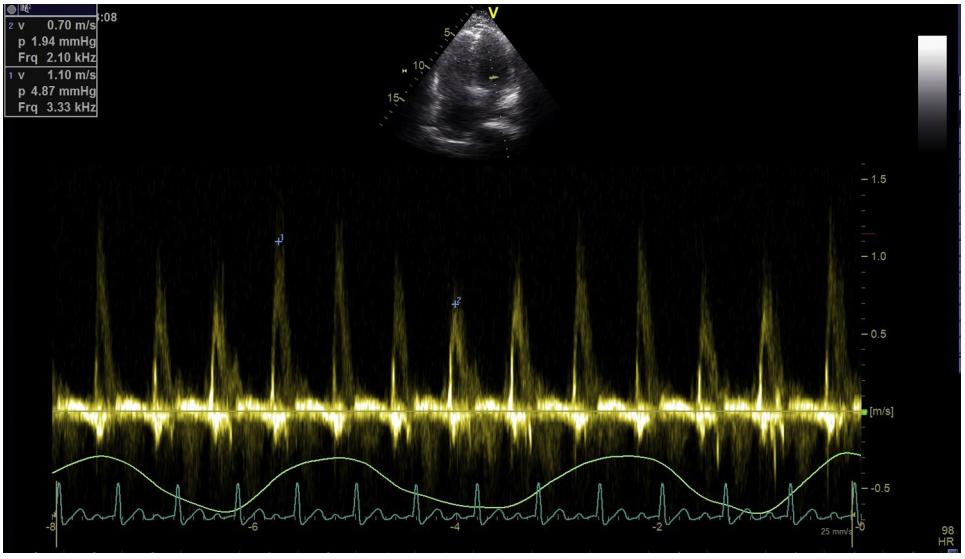

Figure 4 Transthoracic echocardiography (spectral Doppler). Accentuated respiratory variation of mitral flow velocity at 36.6%, an increase from the normal limit of 25%.

Histopathological examination in our case supported the diagnosis of pseudoaneurysm as the hematoma-containing sac was entirely devoid of normal vessel wall architecture as opposed to "true" aneurysm, which consists of intact but weakened wall layers. Although most pseudoaneurysms require inceptive triggers to vascular wall damage for hematoma formation, we are unable to ascertain an attributable origin based on the presentation and investigations. Instead of declaring its spontaneous occurrence, we would consider the strong possibility of preceding tuberculous aortitis as a viable explanation for pseudoaneurysm development. The infection could have been rendered inactive by combination antituberculous chemotherapy resulting in negative histological proof. Evidence on the hematogenous spread of extrapulmonary tuberculosis involving the aorta is well established even among immunocompetent individuals.[9] Autopsy

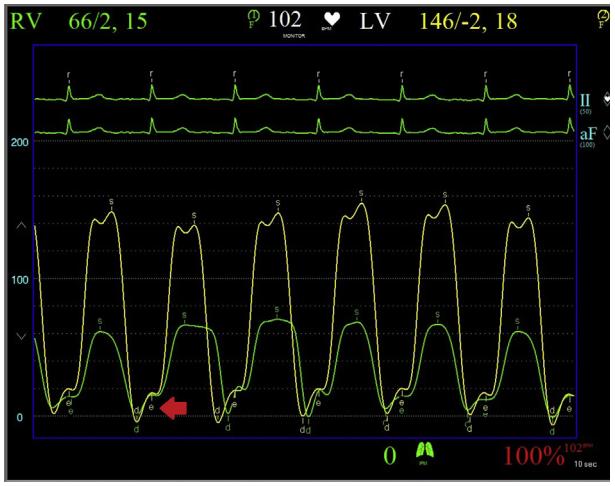

Figure 5 Invasive cardiac hemodynamic study. Left and right ventricles show equalization of pressure tracings during diastole supporting the diagnosis of constrictive pericarditis. Encasement of cardiac chambers results in reduced cardiac compliance and limits the accommodation of blood volume (*green line*: right ventricle; *yellow line*: left ventricle). Impaired myocardial relaxation results in rapid ventricular filling, giving rise to the typical ''dip-and-plateau'' or ''square root'' pattern on hemodynamic tracing (*red arrow*).

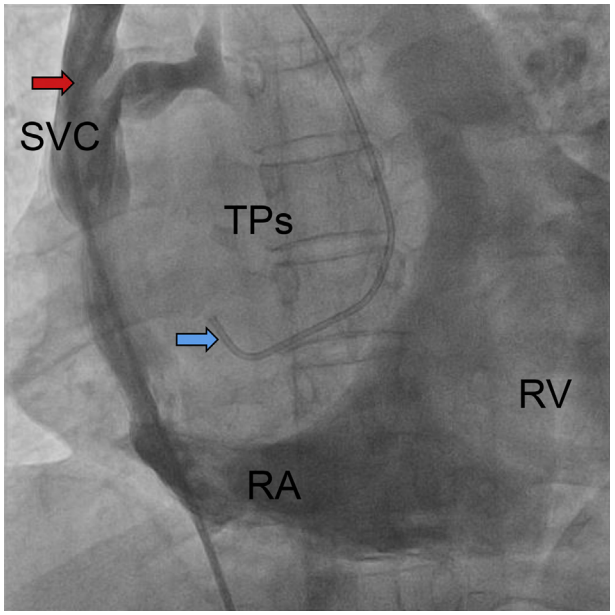

Figure 6 Fluoroscopy imaging during cardiac catheterization. Contrast medium injection from superior vena cava (SVC; *red arrow*) showed deformed and displaced right atrium (RA) and right ventricle (RV) by adjacent thoracic aortic pseudoaneurysm (TPs). The diagnostic catheter (*blue arrow*) marks the silhouette occupied by hematoma sac.

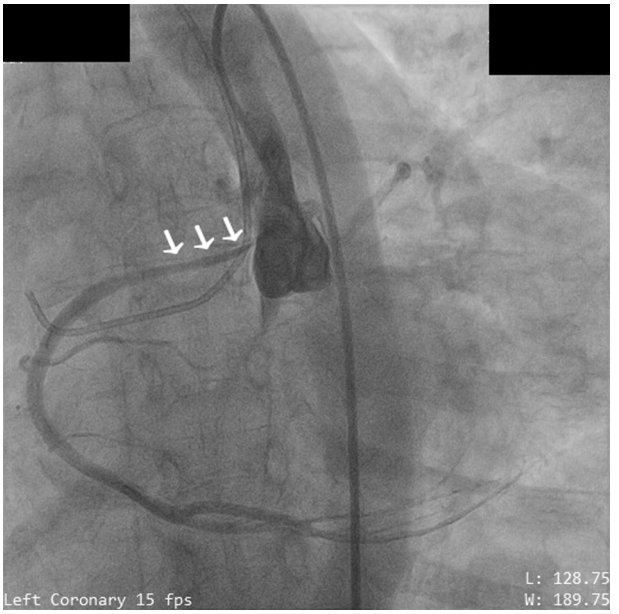

Figure 7 Diagnostic coronary angiography. The right coronary artery is compressed externally and appears elongated at its proximal segment (*white arrows*). The vessel is otherwise free from stenotic disease. Optimal engagement of right coronary ostium is not possible due to deformed aortic root anatomy.

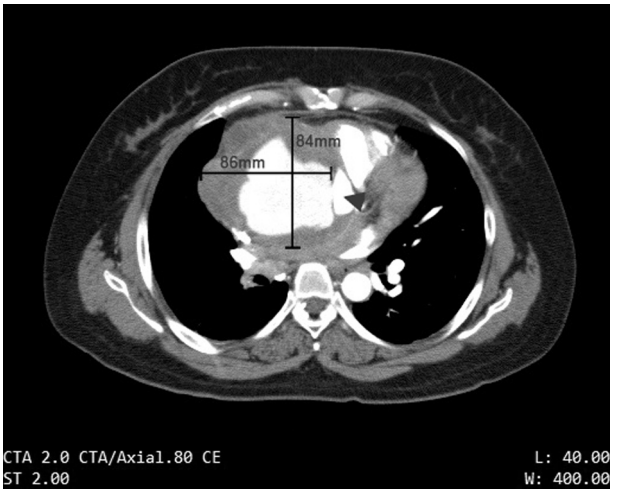

Figure 8 Computed tomography of thorax (axial plane). The entire pseudoaneurysm is fully encased within the pericardium. Dimensions: 84 mm (anterior-posterior, AP) × 86 mm (transverse, TR) × 72 mm (craniocaudal, CC). No evidence of leak or pericardial effusion is seen. The communicating defect of the pseudoaneurysm is indicated (*arrow head*).

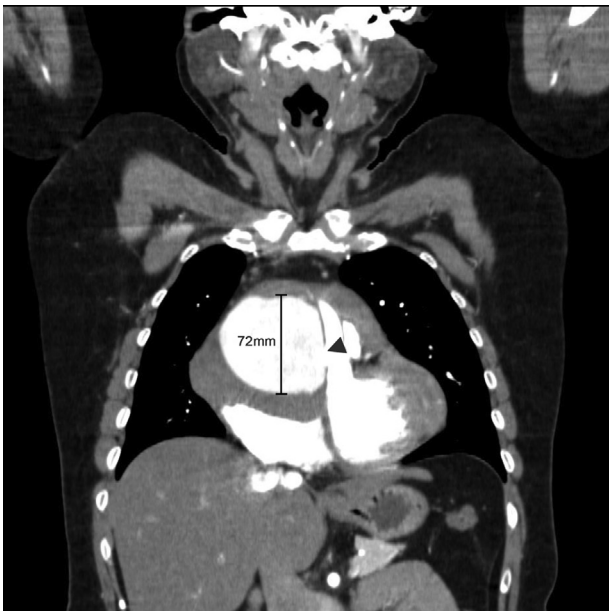

Figure 9 Computed tomography of the thorax (sagittal planes). The neck of the pseudoaneurysm is well-visualized superior to the aortic annulus (*arrow head*). The right atrium and ventricle are caudally displaced by the expansile force exerted by the aneurysmal mass.

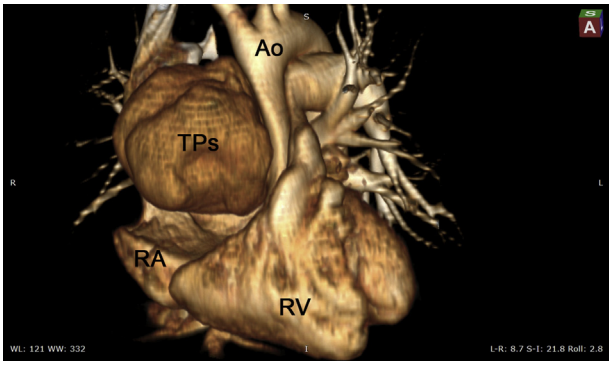

Figure 10 Computed tomography three-dimensional volume rendering imaging (anterior-posterior view). A massive aortic pseudoaneurysm (TPs) compresses a large portion of the ascending aorta (Ao) and caudally displaces the right atrium (RA) and ventricle (RV). *SVC*, Superior vena cava.

findings from a mortality case report demonstrated a causal relationship between tuberculous aortitis and pseudoaneurysm formation due to vascular wall weakening, which unfortunately led to disastrous aortic rupture and death.[10]

CONCLUSION

We reported an unusual case of intrapericardial ascending aortic pseudoaneurysm manifested as constrictive pericarditis. A single unifying

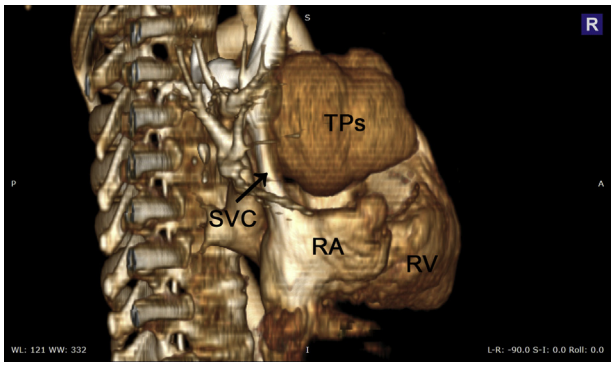

Figure 11 Computed tomography three-dimensional volume rendering imaging (right-lateral view). The superior vena cava (SVC) is squashed and elongated from anterior compression corresponding to the fluoroscopy appearance. *Ao*, Aorta; *RA*, right atrim; *RV*, right ventricle; *TP*, pseudoaneurysm.

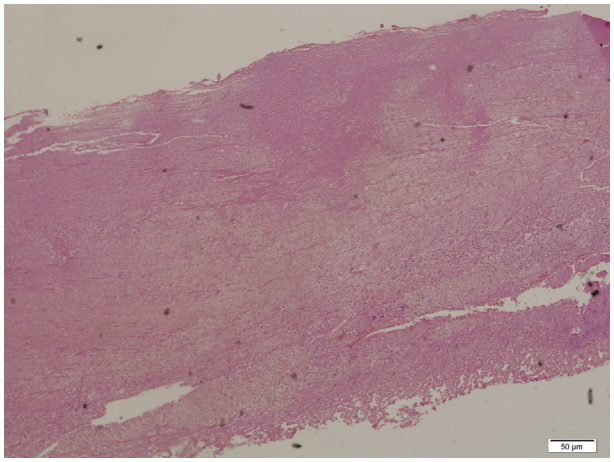

Figure 12 Histopathological examination. The pseudoaneurysm wall showed complete disruption of vessel wall. The section indicates dense fibrous connective tissue devoid of distinct arterial wall layers. Mild to moderate infiltration of neutrophils and lymphoplasmacytic cells was seen. No evidence of granuloma or malignancy.

diagnosis was successfully made based on a multimodality imaging approach.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.04.003.

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