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A Huge Thrombosed Pulmonary Artery Aneurysm without Pulmonary Hypertension in a Patient with Hepatosplenic Schistosomiasis

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 55
Final Diagnosis: Thrombosed pulmonary artery aneurysm
Symptoms: Cough productive • fever • shortness of breath
Medication: —
Clinical Procedure: Pericardiocentesis
Specialty: Cardiology

Objective: Rare disease

Background: We herein report a case of huge pulmonary artery aneurysm in a 55-year-old male farmer from the Nile delta (Lower-Egypt), mostly due to infestation with *Schistosoma mansoni,* which is the parasite causing hepatosplenic schistosomiasis.

Case Report: This male patient was admitted with a month-long history of progressive shortness of breath, 2-month history of fever, and a cough with mucoid sputum for 10 days. On examination, he had normal temperature and blood pressure, but he had tachypnea, tachycardia, and congested neck veins. Electrocardiography showed multifocal atrial tachycardia and right bundle branch block.

Conclusions: The present case is unique in that it shows the presence of a huge pulmonary artery aneurysm despite the absence of pulmonary hypertension.

MeSH Keywords: Aneurysm • Echocardiography, Doppler • Magnetic Resonance Angiography

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/892451

Background

Aneurysms of the main pulmonary artery are rare. An extensive review of proximal pulmonary artery aneurysms (central and peripheral types) by Deterling and Clagett in 1947 [1] documented only 8 cases among 109 571 autopsies spanning a period of 100 years. The etiology and pathogenesis of pulmonary artery aneurysms are not well known. About half are associated with congenital heart disease, the most frequent being patent ductus arteriosus, followed by ventricular and atrial septal defects; most such cases are associated with pulmonary hypertension. Main pulmonary artery aneurysms may also accompany absence of leaflets of the pulmonary valve and stenosis of the ventriculoarterial junction. Pulmonary artery aneurysm is sometimes idiopathic, but some known causes include infection (e.g., syphilis, bacterial endocarditis, tuberculosis), arteriosclerosis, degenerative changes of the elastic media, cystic medial necrosis, trauma, arteriovenous communication, and certain pulmonary vasculitides like Behçet's disease and Hughes-Stovin syndrome [2,3]. Here, we present a case of a huge pulmonary artery aneurysm attributed to schistosomal parasitic infestation, which is endemic in Egypt.

According to the WHO, 200 million people in the world are infected with schistosomiasis [4], which is the third leading parasitic disease in the world after malaria and amoebiasis [5,6]. Also, the incidence of schistosomal cor-pulmonale seen at autopsy in patients with schistosomiasis ranges between 11% and 33% [5,7] secondary to pulmonary hypertension. The present report shows a huge pulmonary artery aneurysm despite the absence of pulmonary hypertension, which is a unique event.

Case Report

A 55-year-old male farmer from the Nile delta (Lower Egypt) was admitted to Mansoura Medicine Specialized Hospital in January 2007 with a month-long history of progressive shortness of breath, 2-month history of fever, and a cough with mucoid sputum for 10 days. He had ceased smoking 5 years previously, and had no history of hypertension, diabetes, tuberculosis, syphilis, autoimmune disorders, blunt or penetrating chest trauma, or malignancy, but reported treatment for intestinal bilharziasis in his 20s.

On examination, he was fully conscious with normal temperature and blood pressure, but he had tachypnea, tachycardia, and congested neck veins. Clinical criteria of Behçet's disease (e.g., oral-genital ulcers and ocular affection) were absent. Cardiac examination results were inconclusive. Electrocardiography showed multifocal atrial tachycardia, right bundle branch block, and right-axis deviation (Figure 1). Chest X-ray revealed

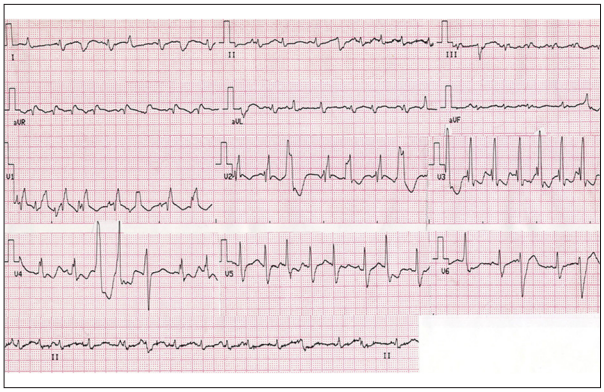


Figure 1. Conventional 12-lead electrocardiogram, showing multifocal atrial tachycardia, right bundle branch block, and right-axis deviation.

cardiomegaly with a dumbbell-shaped heart (Figure 2A, 2B). Arterial blood gas analysis showed PaO_2 76 mmHg, $PaCO_2$ 18 mmHg, SpO_2 93%, and pH 7.66. Serum biochemistry showed normal liver functions, cardiac enzymes, serum creatinine, and full blood picture. Results of investigations for autoimmune disorders (including Behçet's disease) were normal (negative C-ANCA, P-ANCA, ANA-ANF, and normal ESR). Anti-bilharzial-antibody titer (specific for *Schistosoma mansoni*) was 320 (positive >80) and was done twice by 2 different laboratories by using indirect hemagglutination assay (IHA). Abdominal ultrasonography revealed an average-sized liver with cirrhotic echo-pattern and periportal fibrosis, and a moderately-enlarged spleen.

Two-dimensional echocardiography revealed the right side of the heart to be normal, and Doppler study showed normal pulmonary artery pressure (Figure 3A, 3B). Post-contrast chest computed tomography (CT) was done and showed a dilated main pulmonary artery and its 2 branches, with a huge (partially calcified) aneurysm of the right branch (about 17×11 cm) partly occupied by a mural thrombus compressing the left atrium, together with marked pericardial effusion (Figure 4A). A 3D reconstruction of the aneurysm/thrombus complex by CT (Figure 5A, 5B) and magnetic resonance angiography (Figure 5C) was done to confirm the pathologic anatomy.

Pericardiocentesis was undertaken on day 4, immediately followed by subxiphoid drainage, wherein 2500 cc of fluid was aspirated during 1 week. The patient reported improvement of dyspnea. Figure 2 showed chest X-rays done after pericardiocentesis; showing the presence of pericardial air (Figure 2C, arrow) and subxiphoid drainage tube (Figure 2D, arrow) Analysis of the aspirate revealed pure pus (WBCs 4500/cc, RBCs >100/cc, protein 2.8 gm/dl). Two successive cultures of the aspirate, Ziehl-Neelsen stain, adenosine deaminase test, and BACTEC (rapid culture for tuberculosis) were all negative. Pericardial biopsy revealed chronic suppurative inflammation. CT-guided

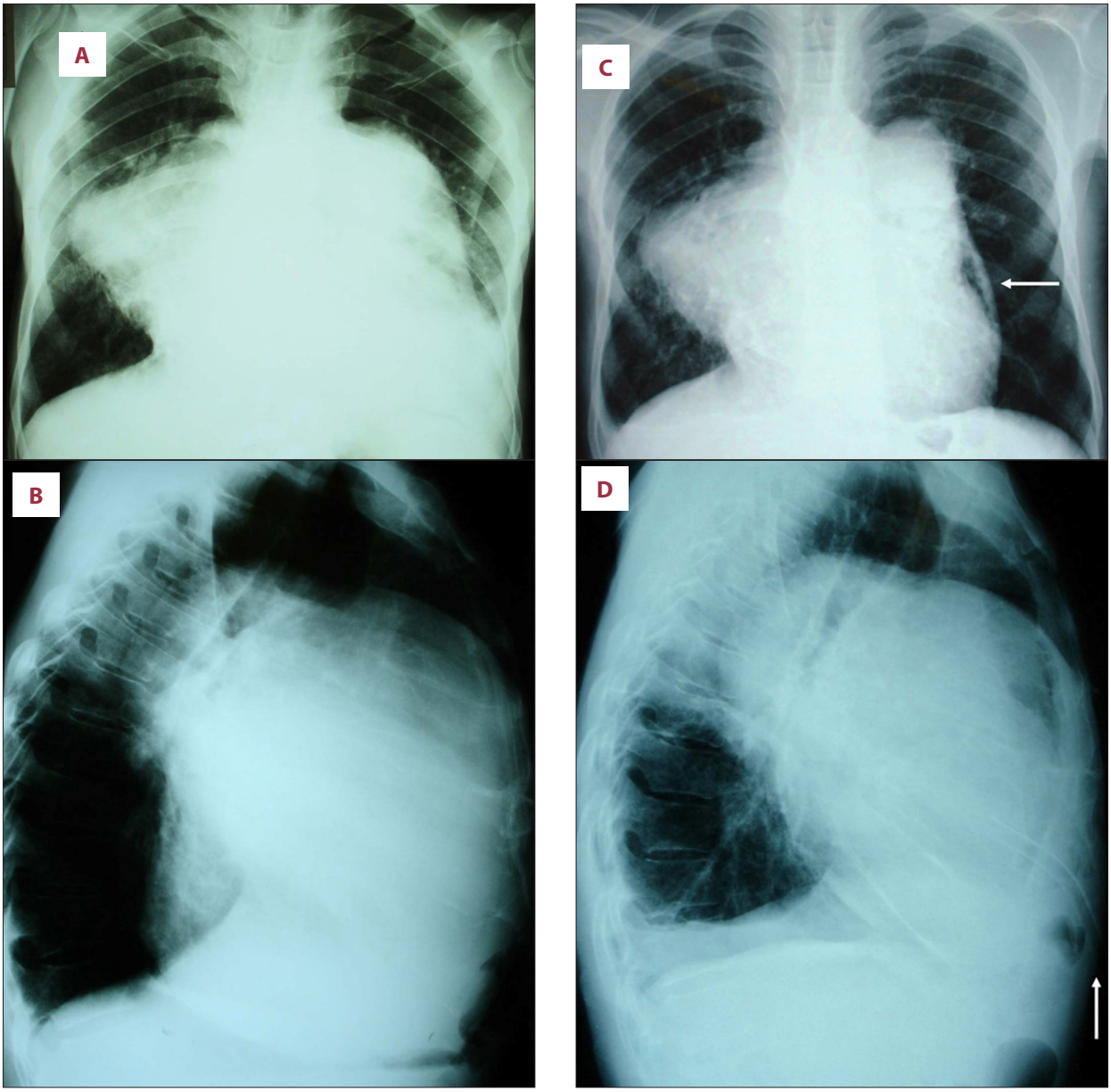


Figure 2. Posterior-anterior (**A, C**) and lateral (**B, D**) chest X-rays before (**A, B**) and after (**C, D**) pericardiocentesis. Note the presence of pericardial air (panel **C**, arrow) and subxiphoid drainage tube (panel **D**, arrow).

needle biopsy was done for a suspected peripheral lung nodule thought to be a malignant lesion, but the histopathologic examination revealed schistosomal granuloma. Surgical correction of the aneurysm with placement of a pulmonary allograft was recommended, but was refused by the patient. Empirical antibiotics, low-molecular-weight heparin, and oral anticoagulant therapies were then instituted.

From day 11 onward, the patient developed recurrent attacks of dyspnea, tachypnea, and tachycardia with stable hemodynamics, suggestive of pulmonary embolic showers. INR was 2.9, but hypoxemia and respiratory alkalosis persisted without evidence of clinical improvement, in parallel with thrombocytopenia (50,000/cc). On follow-up CT (day 17), the thrombus encroached more on the right pulmonary lumen and extended to the left branch, with newly developed right-sided pleural effusion (Figure 4B). Ultrasound-guided aspiration of the pleural fluid (to exclude a leaking aneurysm) revealed transudate. On day 19, the patient suddenly developed marked dyspnea and tachycardia, and passed into shock with marked hypoxemia (PaO_2 65 mmHg) and hypocapnia ($paCO_2$ 17 mmHg), suggesting a massive pulmonary embolism. Then, he went into cardiac arrest (in the form of pulseless electrical activity) and died.

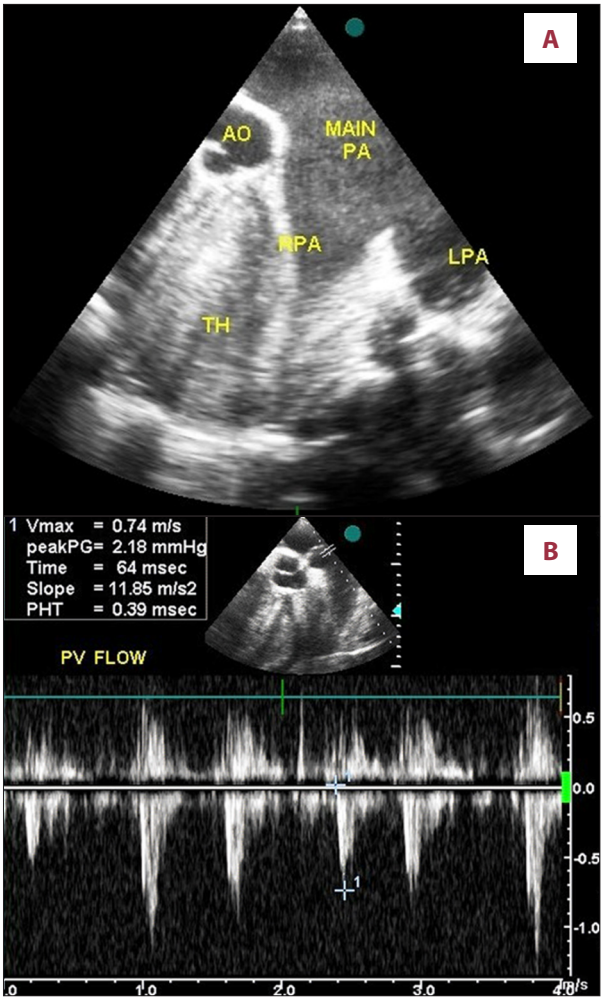


Figure 3. (**A**) Transthoracic 2D echocardiograms showing the right pulmonary artery aneurysm with a contained thrombus. (**B**) Doppler echocardiography revealed normal pulmonary artery pressure. AO: aortic valve; LA: left atrium; LPA: left pulmonary artery; LV: left ventricle; RPA: right pulmonary artery; RV: right ventricle; TH: thrombus.

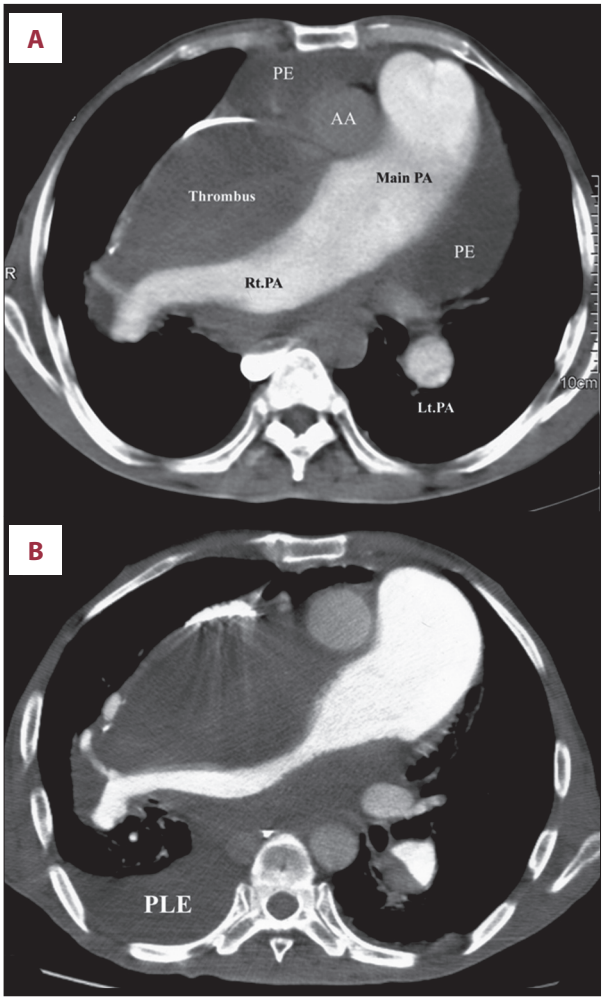
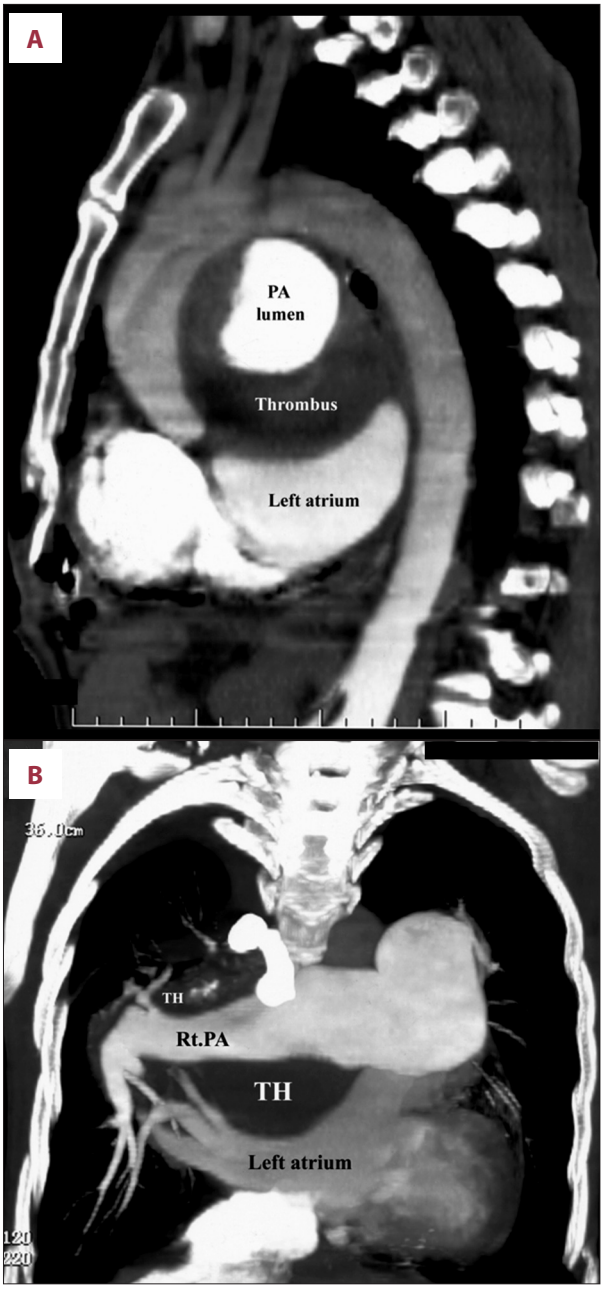


Figure 4. (**A**) Post-contrast chest CT taken on admission, showing a dilated main pulmonary artery (PA) and its 2 branches, with a huge (partially calcified) aneurysm of the right branch (about 17×11 cm) partly occupied by a mural thrombus (TH), together with marked pericardial effusion (PE). (**B**) Follow-up CT showing encroachment of the thrombus on the right pulmonary lumen and its extension to the left branch, with newly developed right-sided pleural effusion (PLE).

Discussion

Pulmonary artery aneurysm is an exceedingly rare finding, but since the first report of schistosomal pulmonary artery aneurysm by Dr. Halim Zaky from Egypt in 1952 [8], another case from Brasil was reported by Piveta et al. in 2012 [9]. Both of these reports directly attribute pulmonary artery aneurysm to *Schistosoma mansoni* parasitic infestation.

Pulmonary artery aneurysm may follow bilharzial cor-pulmonale, which results from *Schistosoma mansoni* infestation that is endemic in Lower-Egypt [10]. The collateral circulation (porto-caval anastomosis) produced by portal hypertension in cases with hepatosplenic schistosomiasis is essential to divert the schistosome and its ova from the portal to the pulmonary vasculature [7,8]. Multiple microembolizations of schistosome eggs produce chronic granulomatous inflammation in and around the pulmonary arterioles, leading to pulmonary hypertension and consequent right ventricular dilatation (cor-pulmonale) [11]. However, the absence of the classic picture of cor-pulmonale in this case and the presence of a chronic huge aneurysm without evidence of pulmonary hypertension is quite peculiar. Most probably, the impact of schistosome-induced damage upon the walls of the main pulmonary arteries was more severe relative to the other segments of the pulmonary circulation, resulting in the formation of pulmonary artery aneurysm. In this respect, obliterative endarteritis in the vasa vasorum of the large pulmonary arteries in cases of pulmonary schistosomiasis has been



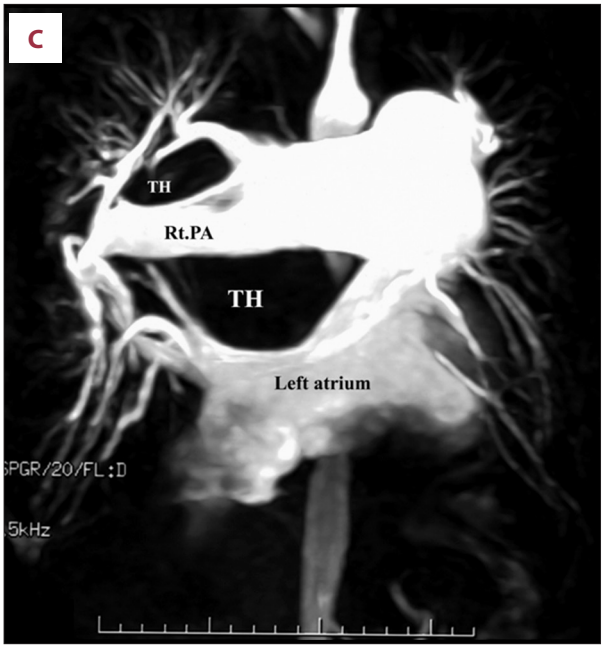


Figure 5. A 3D reconstruction of the aneurysm/thrombus complex by computed tomography (**A, B**) and magnetic-resonance angiography (**C**). Rt.PA: right pulmonary artery; TH: thrombus.

reported to weaken their walls, leading to dilatation even in the absence of pulmonary hypertension [8]. Interestingly, the same mechanism was specified in the first reported case of schistosomal aortic aneurysm, where an extensive infestation of the left upper lobe by ova of Schistosoma mansoni caused extension of the disease into the adjacent pleura and aortic sheath, producing endarteritis obliterans of the vasa vasorum of the aorta and subsequent formation of an aortic aneurysm in the absence of systemic hypertension [12].

Indirect hemagglutination assay (IHA) is a widely used sensitive and specific serological test that can easily detect infection with *Schistosoma mansoni*, and is routinely used in clinical laboratories [13]. Diagnosis of schistosomiasis by detection of specific antibodies is likely to be more sensitive than the traditional method of diagnosis by detection of eggs in stool [14]. Sensitivity of IHA with a cutoff titer of 1:80 ranged from 90.0% to 94.7% and the specificity was 94.7% on average [13]. However, this test is not the only diagnostic tool in this case because of the possibility of a false-positive result (though very low) with IHA, so that abdominal ultrasound was carried out and showed the presence of coarse hepatic periportal (Symmer's pipestem) fibrosis, which is considered pathognomonic for hepatic schistosomiasis [15,16]. Furthermore, the confirmation of schistosomal granuloma on CT-guided needle biopsy left no doubts about schistosomal etiology of this case.

The history of fever and symptoms suggestive of a viral syndrome suggest an attack of acute viral pericarditis complicated by effusion, which accumulated over time and took 1 month to reach to the threshold that could elicit cardiac symptoms. Secondary infection of the pericardial effusion with a pyogenic organism may have taken place, leading to the formation of a purulent exudate.

In this case, thrombophilia was local to the pulmonary aneurysm. The patient gave no history suggestive of thrombosis or embolism in any other part of the body. Also, clinical examination was negative for peripheral thrombosis. Thus, the

disturbed local hemodynamics (turbulent flow) within the aneurysm, the locally destroyed pulmonary arterial intima, and the formation of intimal atheromatosis [7] following the schistosomal reaction, and the subsequent endothelial dysfunction all may have initiated the formation of the intra-pulmonary artery thrombus. The occurrence of purulent infection and dehydration accompanying the untreated febrile episodes most probably precipitated this thrombophilia. Further aggravation and extension of local thrombophilia seemed to occur by the administration of heparin, leading to the well-known complication of heparin-induced thrombotic thrombocytopenia [17], in which massive platelet aggregation happened to take place on top of the previously existing pulmonary artery thrombus, in line with peripheral blood thrombocytopenia [18,19]. Note the reduction of the total platelet count from 140 000/cc to 50 000/cc in parallel with the *de-novo* encroachment of the thrombus on the right pulmonary artery lumen and its extension into the left pulmonary branch, as shown in the follow-up CT.

Conclusions

Though very rare, schistosomiasis should be considered as one of the primary causes of pulmonary artery aneurysm; owing to the presence of huge pulmonary artery aneurysm without pulmonary hypertension in this case.

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