

True aneurysm of the peripheral pulmonary artery due to necrotizing giant cell arteritis

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Abstract

Pulmonary artery aneurysm in adults is a rare diagnosis. Most cases described in the literature are either associated with congenital heart disease or pulmonal arterial hypertension, respectively, or are not true aneurysms but rather pseudoaneurysms, which are usually iatrogenic. We present the case of a 68-year old female patient with the incidental finding of a true aneurysm of the right peripheral pulmonary artery with a maximum diameter of 4 cm. With increasing aneurysm diameter over time, the decision for a surgical resection was made. Complete resection of the aneurysm including lower lobe resection was performed. Histopathological examination showed necrotizing giant cell arteritis as the underlying cause. The postoperative course was uneventful and no signs of further disease activity were detected. To our knowledge, this is the first reported case of a pulmonary artery aneurysm caused by giant cell arteritis, whereas it should be noted that the distinction between Takayasu arteritis and giant cell arteritis is not clearly defined. Considering the high mortality associated with aneurysm rupture, surveillance is advocated for small aneurysms, whereas for larger aneurysms and those showing signs of progression in size despite medical therapy or even dissection, surgical intervention should be considered.

Keywords: Pulmonary artery • Pulmonary artery aneurysm • Giant cell arteritis • Takayasu arteritis

INTRODUCTION

An aneurysm of the pulmonary artery (PAA) is a rare finding. In general PAA can be subdivided into true aneurysms, showing dilatation of all three layers of the vessel wall, and pseudoaneurysms. True PAA are often associated with either congenital heart disease, or are so-called 'acquired', resulting from pulmonary hypertension, inflammation, infection, neoplasm or connective-tissue disease.

What these aneurysms have in common is a certain risk of dissection or even rupture, which often results in imminent death. Because this risk varies with the localization, size and underlying cause, early detection and adequate treatment are crucial in order to prevent high mortality rates.

CASE REPORT

We present the case of a 68-year old female patient who initially presented with symptoms of pneumonia to her family physician. Chest X-ray showed the incidental finding of an unclear circular mass in the right parahilar region. Computed tomography (CT) scan revealed an aneurysm of the right pulmonary artery, distal to the middle lobe arteries, with a maximum diameter of 3 cm. As the patient was completely asymptomatic and echocardiography showed no signs of pulmonary arterial hypertension, the follow-up CT scan was performed with some delay (patient reasons),

after 4 years. As a progression of the aneurysm diameter to 4 cm was documented (Fig. 1), the patient was referred to our department for further treatment. Neither an iatrogenic trauma (i.e. heart catheterization), nor clinical signs of a temporal arteritis could be elicited.

Especially because of the increasing aneurysm size, which now was more than double the normal vessel diameter in this region, the decision for a surgical approach in this asymptomatic patient was made.

On operation, resection and primary reconstruction of the pulmonary artery was not feasible due to the gap in diameter of the vessel in the pre- and postaneurysmatic region, where the aneurysm was expanding into the segmental arteries. Thus, complete resection of the aneurysm including anatomic lower lobe resection was performed (Fig. 1). The postoperative course was uneventful. Histopathological examination showed necrotizing giant cell arteritis (GCA) as the underlying cause for this true PAA (Fig. 2). Taking a potentially generalized disease activity into account, a positron emission tomography was performed in the postoperative course, which did not reveal further foci of inflammation. Since no clinical signs of a temporal arteritis (i.e. headaches, jaw claudication, diplopia or visual loss) were present and erythrocyte sedimentation rates as well as C-reactive protein (CRP) levels were normal in the preoperative period as well as on the follow-up, biopsy of the temporal artery was not indicated. Implying curative surgery, steroid therapy was thus considered abdicable.

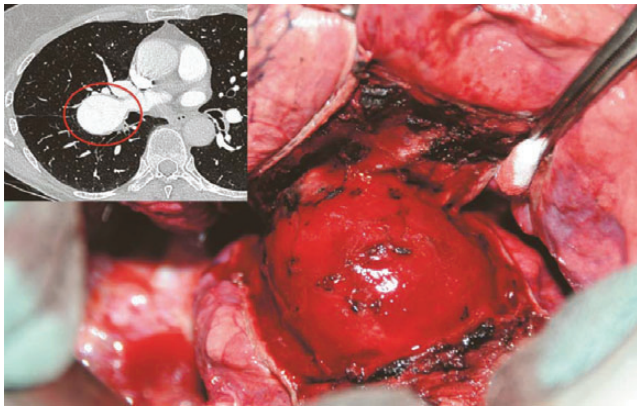


Figure 1: Intraoperative view of the interlobar region with exposure of the pulmonary artery aneurysm. Insert: CT-scan showing aneurysm of the right pulmonary artery measuring 4.1×3.9 cm

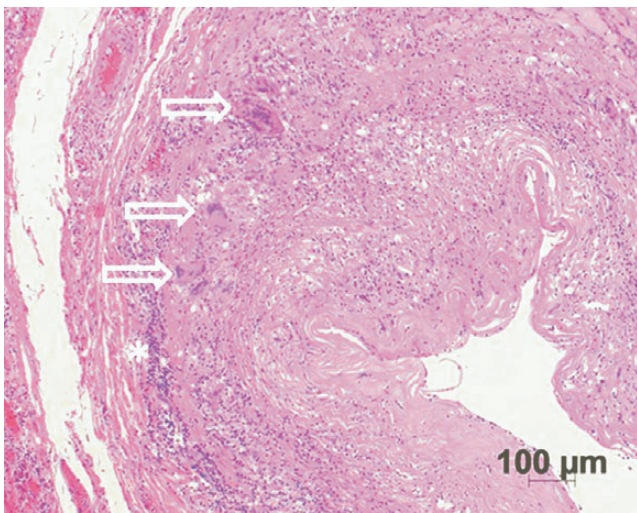


Figure 2: Necrotizing giant cell arteritis of the pulmonary artery: note the giant cells (arrows) and the attendant lymphohistocytic infiltrate (*) with foci of necrosis and fibrosis of the vessel wall (hematoxylin and eosin staining, $10\times$)

DISCUSSION

Of the few reported PAA cases in the literature, $>50\%$ are described as being associated with congenital heart disease, including patent ductus arteriosus, ventricular septal defect or bicuspid aortic valve. However, the sequelae of surgical repair of the aforementioned and other congenital heart pathologies can also be the cause of PAA. In the other half of cases, acquired pathologies are found to be the underlying cause of disease. These include pulmonary arterial hypertension, vasculitis (i.e. Behçet's disease), infection, neoplasm, connective tissue disease (i.e. Marfan syndrome) or trauma (including iatrogenic injuries) [1, 2]. The clinical signs and symptoms are inconsistent and non-specific, since patients may not be symptomatic at all or present with dyspnoea on exertion, cough, chest pain or even haemoptysis. On occasion, the symptoms of a pulmonary embolism (PE) may finally result in the diagnosis of PAA, associated with PE. It is assumed, that PAA also underlie Laplace's law in the lower pressure pulmonary circuit, with haemodynamic forces (i.e. pulmonary hypertension)

promoting PAA-diameter progression, and thus increasing the risk of dissection or rupture [1]. Inayama *et al.* [2] highlighted a mortality rate for PAA of 86.5%, as a rupture of the pulmonary artery wall causes haemopericardium or bleeding into the pleural cavity, leading to a sudden death in most cases. However, because of the rarity of PAA and the diversity of underlying causes, no standardized clinical management and treatment guidelines for PAA have been established yet.

In the presented case, even though clinical manifestations of any infection or inflammatory process were absent and preoperative CRP-levels were not elevated, histopathological analysis revealed necrotizing GCA to be the cause of the aneurysm. For Takayasu arteritis (TA), only a few cases involving the pulmonary artery have been described, but this is the first report of an aneurysm of the peripheral PA due to GCA [3].

Whereas TA predominantly affects the aorta and its major branches in young women, GCA is primarily a disease of the elderly, affecting middle- to small-sized arteries. Inflammatory infiltrates of lymphocytes, giant cells and fibrosis can be found in both diseases, but large numbers of giant cells are typically found in CGA.

Nevertheless, it should be noted that both diseases do overlap, concerning clinical and histopathological findings, and that only a combination of both aspects can lead to diagnosis. Some authors even suggest that both diseases may not be distinct entities, but rather represent different phenotypes of the same disorder [4]. In our case, the localization of the disease, histopathology and the patients' age make GCA the most probable diagnosis, but management options would have been the same for both pathologies.

In PAA, early diagnosis and treatment are crucial as only few cases of surviving patients with ruptured pulmonary artery aneurysms are described [2]. A cut-off in diameter, indicating an increasing risk for severe complications, such as dissection or rupture, has not been defined yet. As a reference, Fraser *et al.* [5] described the upper limit of the normal diameter of the main pulmonary artery to be ~ 29 mm and for the right interlobar artery, ~ 17 mm.

In our opinion, active surveillance by annual CT scans is adequate for small aneurysms, whereas for larger aneurysms (exceeding double the size of the normal vessel diameter) and aneurysms progressing in size despite medical therapy (i.e. corticosteroids), or even dissection, surgical intervention is inevitable. In these cases, surgery appears to be the treatment of choice, being diagnostic and curative.

Conflict of interest: none declared.

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