Main pulmonary artery aneurysm: a case report and review of the literature

Massimo Imazio, Enrico Cecchi, Massimo Giammaria, Franco Pomari, Maria Donata Tabasso*, Aldo Ghisio, Pier Giuseppe Forsennati**, Federico Beqaraj, Elisa Favro, Rita Trinchero, Antonio Brusca***

*Cardiology Department, *Radiology Department, Maria Vittoria Hospital, **University Cardiac Surgery, Molinette Hospital, ***Cardiology Medical School, Turin, Italy*

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Main pulmonary artery aneurysms are a rare entity with few available published data. As reported in the literature, operative treatment is commonly recommended but the relation between the size of the aneurysm, its localization, and the risk of rupture is not as well defined as for aortic aneurysms. Proximal lesions that involve the main branches of the pulmonary artery are usually apparent on chest radiographs and must be taken into consideration in the differential diagnosis of mediastinal masses. An early diagnosis allows timely surgical treatment.

We report an unusual case of a main pulmonary artery aneurysm presenting with persistent non-productive cough and provide a review of the pertinent published data.

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Address:
Dr. Massimo Imazio
Corso Trapani, 195/A
10141 Torino
E-mail: imazio@tin.it

Introduction

Main pulmonary artery aneurysms (PAAs) are rare with few reported cases. The natural history is not completely understood and no clear guidelines about the surgical indications are available (the relation between the size of the PAA, its localization, and the risk of rupture is not as well defined as for aortic aneurysms).

We report an unusual case of a main PAA due to isolated cystic medionecrosis presenting with persistent, non-productive cough. Cough is a rare, non-specific symptom in this disease and PAA was a casual finding during diagnostic evaluation.

Case report

A 64-year-old male patient was admitted to the emergency department because of intense non-productive cough. The patient had no history of ischemic heart disease, cardiomyopathy, pulmonary and systemic disease, chest trauma, and cigarette smoking. Physical and neurologic evaluations were normal. Blood pressure was 120/80 mmHg. Blood tests were all within normal range as well as the basal ECG (Fig. 1). Chest radiography (Fig. 2) and transthoracic echocardiography showed a grossly enlarged main pulmonary artery (Fig. 3). No previous chest radiographs were available. Mild pulmonary valve insufficiency was detected but the right atrium and ventricle were normal. Pulmonary systolic pressure, estimated at Doppler echocardiography, was normal (Fig. 4) and no other structural abnormality was found. Transesophageal echocardiography (Fig. 5) and chest computed tomography (Fig. 6) confirmed the presence of a main PAA with a diameter of 6 cm. Screening for rheumatic and autoimmune diseases as well as diagnostic evaluation for tuberculosis, syphilis and the Behçet and Marfan syndromes were negative. Coronary angiography was normal, while pulmonary angiography confirmed the presence of a main PAA (Fig. 7).

The size of the aneurysm and a literature review induced us to propose surgical therapy to the patient. An aneurysmorrhaphy was performed with a satisfactory outcome. At surgery, the pulmonary valve appeared normal and there was no evidence of intimal tear or medial dissection of the pulmonary artery (Fig. 8). Pathologic examination of the aneurysmal wall showed cystic medionecrosis. At 18 months of follow-up the patient is alive and free of symptoms.

The exact mechanism of the PAA in this patient is unclear; however, a structural weakness of the pulmonary arterial wall

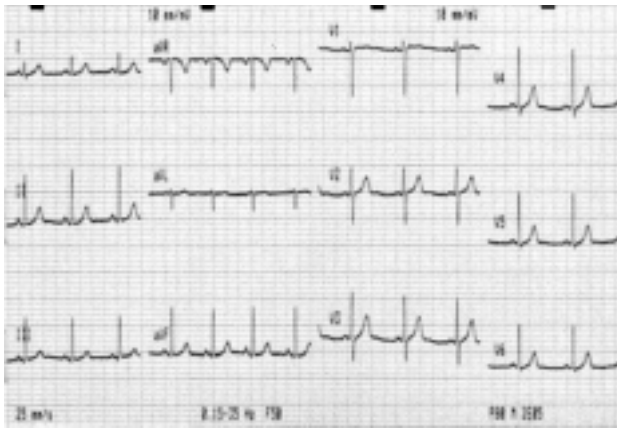


Figure 1. *The standard 12-lead electrocardiogram was normal.*

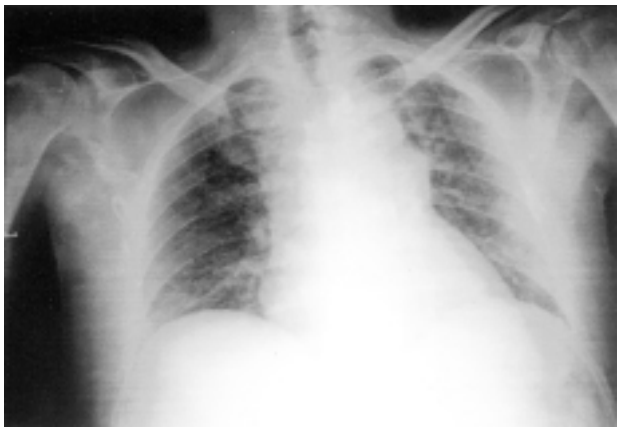
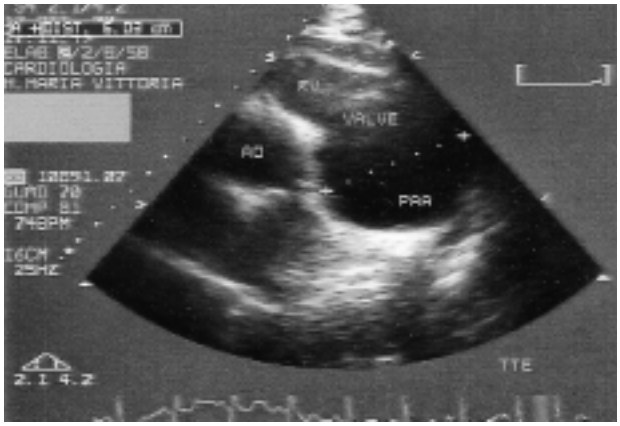


Figure 2. *Postero-anterior chest roentgenogram showing a grossly enlarged main pulmonary artery.*



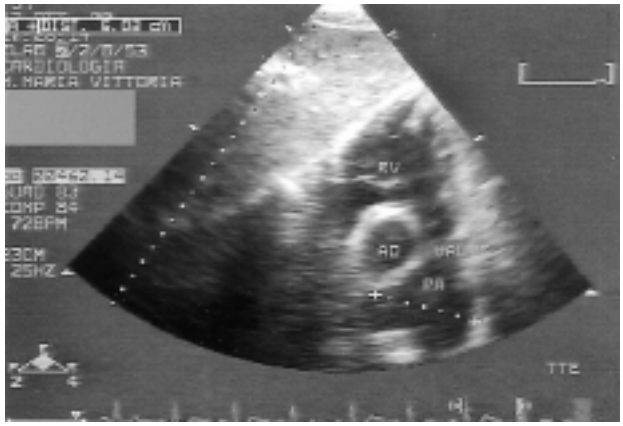
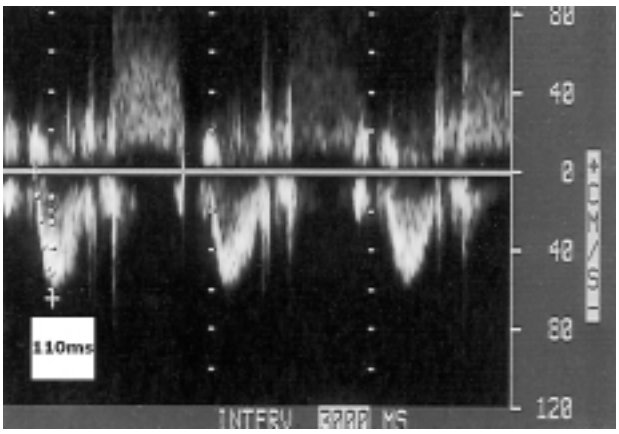


Figure 3. *Transthoracic echocardiography (TTE): a pulmonary artery aneurysm (PAA), approximately 6 cm in diameter, is shown in the parasternal short-axis view at the level of the aorta (AO) and main pulmonary trunk* (left panel) *and in the subcostal short-axis view of the right ventricular outflow tract* (right panel)*. PA = pulmonary artery; RV = right ventricle.*



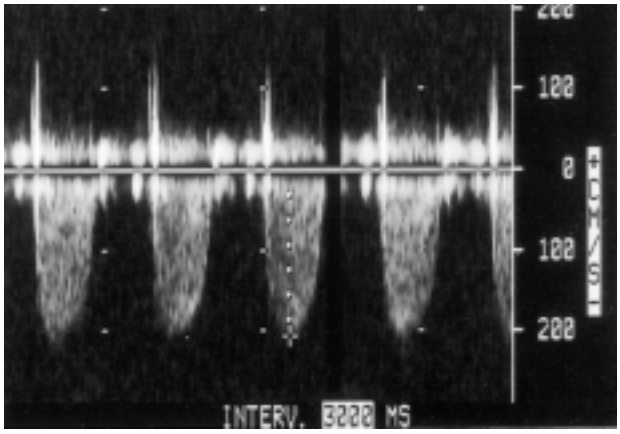


Figure 4. *Doppler findings: a right ventricular acceleration time of 110 ms (normal) was obtained using pulsed wave Doppler with the sample placed within the right ventricular outflow tract* (left panel)*; the Doppler signal of the tricuspid regurgitant jet was obtained by continuous wave Doppler from the apical 4-chamber view; the peak velocity measured approximately 2.0 m/s giving an estimated systolic pulmonary pressure of 26 mmHg (assuming that the right atrial pressure equals 10 mmHg;* right panel).

may be a possible explanation for the development and progression of the PAA. The presence of symptoms and the potential risk of enlargement and rupture were the rationale for proposing surgical therapy in this case.

Discussion

PAA is a rare disease with an estimated autoptic incidence of about 8 cases every 100 000 autopsies[1,2].

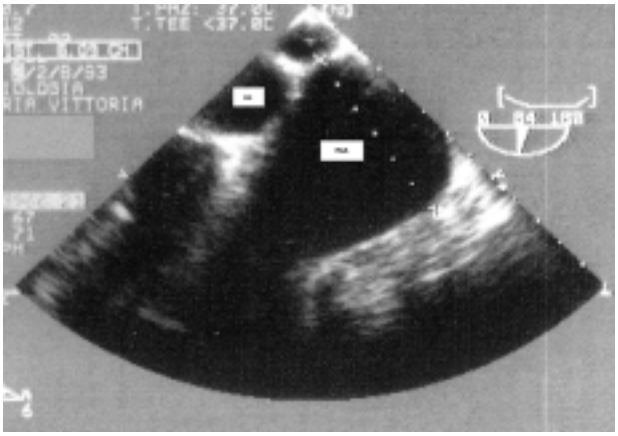


Figure 5. *Transesophageal echocardiography: a multiplane transesophageal view at 84° obtained from the upper esophageal position. It shows a pulmonary artery aneurysm (PAA) with a diameter of approximately 6 cm. Ao = aorta.*

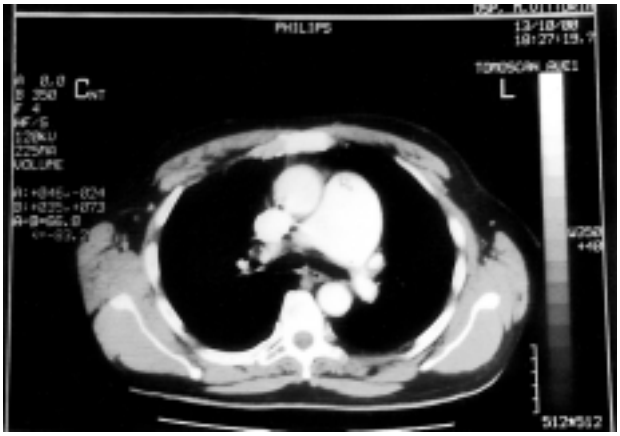


Figure 6. *Contrast-enhanced computed tomography confirmed the presence of a main pulmonary artery aneurysm with a diameter of approximately 6 cm* (white arrow).

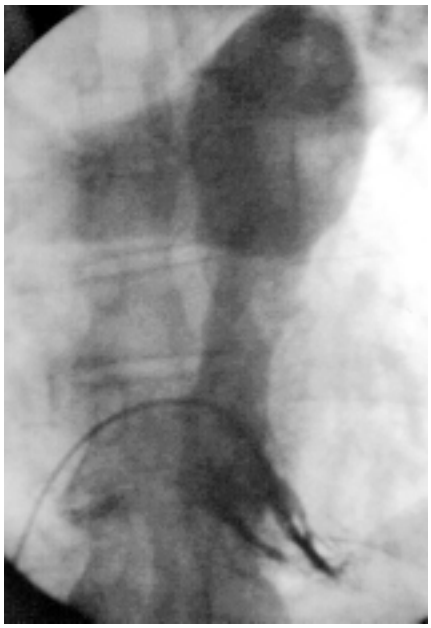


Figure 7. *Catheterization image of the pulmonary artery aneurysm (postero-anterior projection).*

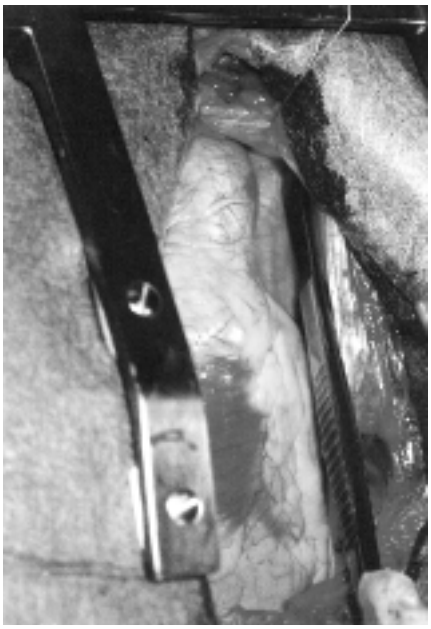


Figure 8. *Surgical view showing the pulmonary artery aneurysm originating from the right ventricle.*

A proposed classification of PAAs[1] divided these entities into those with and those without an arterio-venous communication (Table I).

In the preantibiotic era, tuberculosis and syphilis caused the majority of cases. Syphilitic PAAs were typically proximal because vessel wall weakening began with vasa vasorum infection, while tuberculous aneurysms were distal involving the small intraparenchymal arteries[1].

Although the etiology and pathogenesis of PAA are not well known, nowadays a simplified and practical classification for the management of PAA may only include primary PAAs and secondary PAAs. Huge PAAs without pulmonary hypertension are commonly called "low pressure giant PAAs"[3].

Table I. Classification of pulmonary artery aneurysms (PAAs).

PAAs without an arterio-venous communication
Infection (mycotic aneurysms): tuberculosis (Rasmussen's aneurysm), syphilitic, other (bacterial, fungal)
Structural heart disease (congenital heart disease, acquired cardiac disease)
Structural vascular disease (Marfan syndrome, Behçet syndrome, vasculitis)
Pulmonary hypertension
Idiopathic (isolated, Hughes-Stovin syndrome)
Trauma
PAAs with an arterio-venous communication
Congenital (isolated, associated with hereditary hemorrhagic teleangectasia)
Acquired (infection, trauma)

Approximately half are associated with congenital heart disease, the most frequent being patent ductus arteriosus, followed by ventricular and atrial septal defects and transposition of the great arteries with ventricular septal defect[1,3-6]. Many cases are associated with pulmonary hypertension. Other causes include infections that are less common than in the past. Tuberculosis was the first identified cause of PAA and a tuberculous PAA was first described by Rasmussen. The aneurysm described by this author was due to an extension of the infection from the surrounding lung tissue involving the intrapulmonary vessels. PAA generally occurred in patients with chronic progressive disease with an autopsy prevalence of 4 to 11%. Due to immigration, the increasing frequency of tuberculosis in developed countries makes this finding a topical subject even nowadays[7,8].

Several bacteria and fungi have been implicated in less frequent forms of mycotic PAA including *Staphylococcus aureus*, *Streptococcus sp, Corynebacterium diphtheriae, Candida albicans* and *Aspergillus flavus*. PAAs were described in association with infective endocarditis[9], skin abscesses, and pneumonias[10].

More recently, associated connective tissue disorders such as Marfan syndrome and Ehlers-Danlos syndrome as well as systemic vasculitides such as Behçet syndrome have been identified as causes of PAA[1]. The vasculitis of Behçet syndrome is now a well-known cause of aneurysms of the main pulmonary arteries and their branches[11-13].

Extravascular or intravascular trauma is a rarely documented cause of PAA, but may more commonly be a cause of pulmonary artery pseudoaneurysm. An increasing number of cases are iatrogenic and due to the use of the Swan-Ganz catheter[14,15].

Some idiopathic cases of PAA may be associated with the Hughes-Stovin syndrome, a rare syndrome with recurrent superficial and deep venous thrombosis, an increased intracerebral pressure (due to venous thrombosis) and PAA[1,16].

The clinical manifestations of PAA are often nonspecific and possible symptoms include hemoptysis, dyspnea, chest pain, and cough. Hemoptysis is an important symptom because it is a marker of instability and a strong indicator of the need for intervention[1].

The main diagnostic testing modalities for PAA are chest radiography, echocardiography, contrast-enhanced computed tomography, magnetic resonance imaging, and pulmonary angiography. Though angiography has been considered the gold standard for the diagnosis of PAA[17,18], the diagnosis of this entity usually requires the combination of multiple imaging modalities. Magnetic resonance imaging has emerged as a useful non-invasive imaging modality that is ideal for the detection of possible intimal flaps and for the long-term monitoring of the size of the aneurysm[18].

The natural history of PAA is not completely clear: few surveys have been reported in the literature and probably many cases, above all asymptomatic cases, are ignored. Some estimate that up to one third of patients with PAA die of rupture[19,20] but some aneurysms have persisted for 3 to 17 years without rupture[21]. Possible complications of PAA include dissection, intrapulmonary erosion, pulmonary embolism and compression of the trachea, bronchi, superior vena cava or recurrent laryngeal nerve.

Rupture and dissection are the most dreadful complications. Although they are rare, these complications are life-threatening events. Since the first report of a dissecting aneurysm of the pulmonary artery in 1862, 48 cases have been reported[22,23]. Dissection mostly develops in PAA associated with pulmonary hypertension and/or connective tissue disease[22] while reported risk factors for rupture are advanced age and the previous use of steroids[24]. In pulmonary artery dissection, the false lumen tends to rupture rather than to develop a reentry site, as is usual in aortic dissection.

Any PAA, particularly when associated with hemoptysis, cannot be considered to be stable. No controlled studies are available and only case histories are useful for prognostic considerations. So far, collected case histories are strongly in favor of intervention[1,19-21,25,26].

On the contrary, some authors reported a possible conservative management strategy for the so-called "low pressure giant PAA"[3]. On the basis of previous reports which documented progression to arterial wall dissection and rupture mainly in patients with pulmonary artery hypertension, associated congenital heart disease or connective tissue disorders, these authors have concluded that in low pressure giant PAA the risk of rupture seems very small and that conservative management may be useful[3,6,27,28].

Unfortunately, although rare, pulmonary artery dissection has been documented even without underlying pulmonary hypertension or a definite cause[22,29]. Moreover, progressive enlargement of a proximal PAA has been described after 4 years of follow-up even in the absence of an intracardiac shunt and pulmonary hypertension[30]. Thus, decision-making remains difficult and controversial even in the individual patient with a low pressure giant PAA.

The management of PAA has largely been surgical. Reported procedures have included aneurysmectomy with Dacron graft placement or autologous pericardial replacement or arterial aneurysmorrhaphy (Table II)[3,18,22,25,26,31-33]. The first successful aneurysmectomy was performed in 1971[32]. Aneurysmorrhaphy is a simple and quick procedure but with possible recurrent pulmonary dilation due to incomplete resection of the aneurysmal wall especially in cases with associated pulmonary hypertension or structural heart disease. More peripheral lesions may be treated with aneurysmectomy, ligation, segmental resection, lobectomy, pneumonectomy or non-surgical embolotherapy.

In conclusion, no clear guidelines about the surgical indications are available. Main PAAs probably do not

Table II. Literature review of the main reported surgical series and cases with main pulmonary artery aneurysm (PAA) after 1990.

Authors	Age (years)	Sex	PAA size (mm)	Associated conditions	Operation
Chen et al.[25], 1996	55	M	160 × 90	PS	Aneurysmectomy
Roth et al.[26], 1999	65	M	72	PI, CAD	Aneurysmectomy
Kuwaki et al.[31], 2000	54	F	62	PS	Aneurysmorrhaphy
Kuwaki et al.[31], 2000	55	F	73	MS	Aneurysmectomy
Kuwaki et al.[31], 2000	66	M	72	PS, DA IIIb, AAA	Aneurysmectomy
Kuwaki et al.[31], 2000	78	F	60	PS, AR, DA IIIa, CAD	Aneurysmorrhaphy
Senbaklavaci et al.[22], 2001	34	F	80	PPH	Aneurysmectomy
Nair and Cobanoglu[18], 2001	63	M	65	None	Aneurysmorrhaphy
Veldtman et al.[3], 2003	59	F	90	PI, PDA	Aneurysmectomy
Veldtman et al.[3], 2003	37	M	80	PI	Aneurysmorrhaphy
Veldtman et al.[3], 2003	64	F	70	PI	Aneurysmorrhaphy
Veldtman et al.[3], 2003	45	M	100	PI	Aneurysmorrhaphy

AAA = abdominal aortic aneurysm; AR = aortic regurgitation; CAD = coronary artery disease; DA = dissecting aortic aneurysm; MS = mitral stenosis; PDA = persistent ductus arteriosus; PI = pulmonary insufficiency; PPH = primary pulmonary hypertension; PS = pulmonary stenosis.

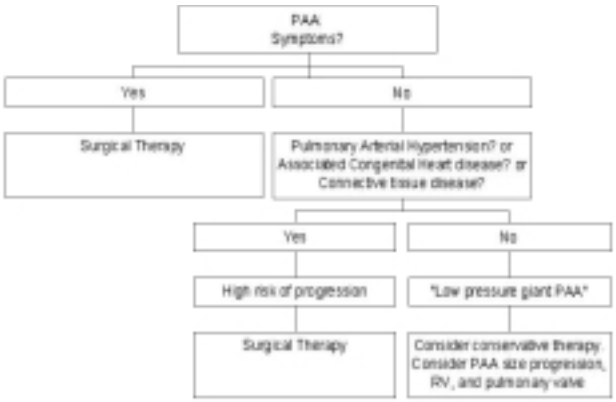


Figure 9. *A proposed management strategy for pulmonary artery aneurysm (PAA). RV = right ventricle.*

warrant the same aggression as aneurysms of the aorta do, especially if the pulmonary artery pressure is normal. However, aneurysmal rupture frequently manifests as sudden death and may occur even in the absence of pulmonary hypertension. Symptomatic cases, patients with pulmonary arterial hypertension or associated congenital heart disease as well as connective tissue disorders have a documented increased risk of progression to dissection or rupture and are candidates for surgery[1,3,6,18,27,28,31]. In the opinion of several authors, when the PAA involves the main pulmonary trunk surgical intervention is generally indicated, to prevent a possible fatal aneurysm rupture, if the patient has an acceptably low operative risk regardless of the etiology and underlying disease[31]. Patients with the so-called "low pressure giant PAA" may have a lower risk of progression and a conservative management strategy may be advisable. In these cases, the timing of surgical intervention should be determined on the basis of the progressive changes in the size of the PAA and/or in the right ventricular size and function[3,6,18] (Fig. 9).

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