Aneurysm of the pulmonary artery with cystic medial necrosis and massive pulmonary valvular insufficiency

Report of two successful surgical cases

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Abstract. The authors report two cases of true aneurysms of the pulmonary artery (APA) associated with massive pulmonary valvular insufficiency. Both were associated with pulmonary artery hypertension and showed cystic medial necrosis at microscopic examination. One case had no associated defects while the other had a large ventricular septal defect and a small patent ductus arteriosus. Both were treated by aneurysmorhaphy and a valvular procedure (valve replacement, valvuloplasty). The clinical results were excellent. Catheterization at 2 years showed no further aneurysm formation, no pulmonary hypertension, and mild pulmonary valve insufficiency in both patients. The literature concerning APA is reviewed and the etiology and the surgical treatment of APA discussed. A unique method of treating pulmonary valve insufficiency is reported.

Key words: Aneurysm – Pulmonary artery – Valvular insufficiency

Aneurysms of the pulmonary artery (APA) are rare. The dilatation is usually limited to the main trunk, and there are histologic degenerative changes of cystic medial necrosis in the wall [3]. These aneurysms are often associated with intra-cardiac defects which have increased pulmonary blood flow leading to pulmonary artery hypertension (PAH), although some cases of APA without PAH [13] or intracardiac defects [10, 15] have been described.

Severe pulmonary valve insufficiency associated with an APA has rarely been reported. The purpose of this paper is to report two cases of APA with massive pulmonary valve insufficiency and cystic medial necrosis. In both cases there was PAH. In one case there was an associated large ventricular septal defect (VSD) and a small patent ductus arteriosus (PDA).

Both patients underwent successful surgical repair. One had aneurysmorhaphy, closure of the VSD and the PDA, and pulmonary valve replacement. A commissuroplasty was performed on the other. Both patients were recatheterized 2 years after surgery. There was no progression of aneurysm, no PAH, and trivial pulmonary valve insufficiency in both.

Case reports

Case 1

A 27-year-old African male had increasing exertional dyspnea for 4 years. There was no history of syphilis or of chest trauma. On admission, he was in good clinical condition, NYHA functional class II, with atypical left-sided chest pain.

On examination, there was no sign of cardiac failure. The left ventricular heave was displaced leftwards. The pulses were regular, the blood pressure was 140/80. There was a grade II/VI systolic murmur heard along the left sternal border in the third and fourth costal interspaces and a grade IV/VI diastolic murmur with a diastolic thrill in the same area. Except for a slight splenomegaly, the remainder of the examination was unremarkable.

The chest X-ray film (Fig. 1) showed moderate heart enlargement and an aneurysmal dilatation of the main pulmonary artery. The right pulmonary artery (RPA) and the left pulmonary artery (LPA) were also dilated in their hilar portion. The ECG showed right axis deviation and right ventricular hypertrophy. The echocardiogram showed a dilated main pulmonary artery (MPA). The semilunar valves could not be clearly identified.

Cardiac catheterization (Table 1) showed moderate pulmonary artery hypertension and no detectable shunts. There
Fig. 1a, b. Case 1: a Preoperative AP chest X-ray film showing an aneurysm of the main pulmonary artery branches. b Two years postoperatively, the main pulmonary artery is almost normal (arrows). The diameters of the branches are slightly decreased as compared to preoperatively.

Fig. 2a, b. Case 1: a Preoperative; b postoperative (2 years). Lateral right ventricular angiograms showing the size of the main pulmonary artery and its late postoperative decrement. In b, the pulmonary valve leaflets are clearly visualized.
widening of the aneurysm, which began at the level of the pulmonary insufficiency (Fig. 2).

There was no systolic gradient across the right ventricular outflow tract. The annulus was not dilated, and the valvular wall was abnormally thickened.

Pass (CPB) with hemodilution and moderate hypothermia. The anterior wall of the aneurysm, including the major part of the pulmonary annulus. At surgery, the aneurysm had a presumed etiology of the pulmonary insufficiency was a pulmonary valvular agenesis. At surgery, the aneurysm had a size-29 bioprosthesis. A large anterior portion of the aneurysm (15 cm diameter) was opened longitudinally. The arterial wall was abnormally thickened.

The pulmonary valvular insufficiency seemed to be the result of a separation of the commissures due to the proximal widening of the aneurysm, which began at the level of the annulus. The annulus was not dilated, and the valvular structure appeared normal. There was no subvalvular stenosis and no ventricular septal defect. A wide resection of the anterior wall of the aneurysm, including the major part of the anterior valvular sinus, was performed so that after direct sutures of the edges of the resection, two commissures were brought together and nearly normal valvular competence was restored (Fig. 3). The postoperative course was uneventful, and the patient was discharged with a grade I/VI soft diastolic murmur along the left sternal border.

Histological examination of the arterial wall showed a thickening of the endothelium without lipidic deposits or calcifications. There was typical cystic medial necrosis in the arterial wall. Two years later, the patient was NYHA class I and off all medication. There was a grade II/VI systolic murmur and a grade II/VI diastolic murmur along the left sternal border.

The chest X-ray film was similar to the early postoperative findings with dilatation of the proximal right and left pulmonary arteries (slightly decreased compared with preoperative X-ray films) and an almost normal-sized pulmonary artery (PA) trunk (Fig. 1). The patient was catheterized (Table I), and there was no PAH noted. The angiogram showed the pulmonary valve to be competent, and the main PA was considerably smaller than preoperatively (Fig. 2).

**Case 2**

An 18-year-old African male was admitted with an aneurysm of the pulmonary artery and a VSD. He had dyspnea on exertion since childhood but was first examined medically at the age of 16, when he presented with an episode of congestive heart failure.

On admission, he was in good clinical condition, NYHA class II. His blood pressure was 120/80, and his pulses were normal. Examination of the chest showed the point of maximal impulse above the left nipple, with an intense systolic and diastolic thrill. Auscultation revealed a grade IV/VI systolic murmur and a grade IV/VI diastolic murmur heard maximally along the left sternal border.

The chest X-ray film showed cardiomegaly with aneurysmal dilatation of the main pulmonary artery. Both right and left pulmonary arteries were dilated proximally (Fig. 4). ECG showed right and left ventricular hypertrophy.

At catheterization (Table I), there was a left-to-right shunt at the ventricular level with a QP/QS of 2.1:1.0 and systemic PAH. There was a low cardiac index (1.9). The angiograms showed a subpulmonary VSD, an enormous aneurysm of the PA, and massive pulmonary valvular insufficiency.

The patient was operated on in November 1982. After establishing cardiopulmonary bypass with hemodilution and using cardioplegia in the aortic root, the aneurysm (15 cm diameter) was opened longitudinally. The wall of the aneurysm was thickened, and the orifices of the RPA and LPA were enlarged. The pulmonary annulus was extremely dilated. The wall of the aneurysm was thickened, and the orifices of the RPA and LPA were enlarged. The pulmonary annulus was extremely dilated, but the valve leaflets seemed to be normal with only slight thickening. There was a small PDA (2 mm) and a large subpulmonary VSD. The PDA was closed with pledgeted sutures. Two of the pulmonary cusps were resected while the one adjacent to the VSD was turned down and used as a patch to close the VSD. The pulmonary valve was replaced by a size-29 bioprosthesis. A large anterior portion of the aneurysm was resected and the vessel closed with a double running suture of 5/0 monofilament. CPB was easily discontinued. There was an early reoperation for bleeding. The further postoperative course was uneventful. The patient was kept on digoxin and diuretics.

Pathologic examination of the arterial wall showed cystic medial necrosis. The pulmonary cusps showed mild thickening and scarring. The patient was followed regularly and, 15 months after surgery, remained in NYHA class I. Examination showed a grade I/VI diastolic murmur along the left sternal border. On chest X-ray film, the cardiomegaly had decreased (Fig. 4).

The patient was subsequently recatheterized. There was no left-to-right shunt, and the pressures in the right ventricle (RV) and PA were nearly normal (34 and 32 mmHg). On angiogram, mild dilatation of the MPA, RPA and LPA

### Table 1. Cardiac catheterization

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<th>Patient</th>
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<tr>
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RAP = right atrial pressure; RVP = right ventricular pressure; PAP = pulmonary artery pressure; CWP = capillary wedge pressure; CI = cardiac index

### Fig. 3a, b. Case 1: Drawing of the operation. a The aneurysm is longitudinally opened. The commissures of the pulmonary valve are "everted", and there is massive pulmonary insufficiency. b After a large aneurysmal resection done deeply in the valvular sinus and after a longitudinal suture of the PA trunk, the valvular competence is restored by approximation of two commissures.
Discussion

Aneurysms of the pulmonary artery have rarely been reported [10]. An exhaustive study of the literature in 1982 [16] showed 300 cases studied and reported. Pulmonary artery aneurysms are most often located in the MPA (89%) and sometimes also in the main branches (11%) [12]. In these cases, the aneurysms are often multiple and sometimes bilateral. In the case of multiple aneurysms, the etiology may be mycotic, a compli-
ication of repeated pulmonary embolism, or unknown. To be considered a true aneurysm, the pathology must include a deformity of the MPA and sometimes of the branches with fusiform or sacciform dilatation and loss of normal architecture in the arterial walls.

The etiology of APA has been discussed extensively [12, 15, 16]. The most commonly reported etiology of APA is association with congenital heart disease with a large left-to-right shunt and PAH.

The most frequently associated defects are PDA and VSD [2, 5, 21]. A true APA should be distinguished from the massive pulmonary artery dilatation (MPA and branches) encountered in the congenital absence of the pulmonary artery valves with or without a VSD and an infundibular stenosis (tetralogy of Fallot with absent pulmonary valves) [26].

Syphilis has been thought to be the major cause of APA in 24% to 39% of the cases [2, 5]. In such cases, the patients are usually young, and the PA is involved sooner than the aorta in the evolution of the disease. A mycotic origin has also been described, sometimes associated with congenital heart disease (CHD) [2, 9]. However, it may be found without CHD [2]. There is bacterial involvement with erosion of the vascular wall and structural alterations of the media.

Atherosclerosis is found in 23% to 30% of these cases [2, 5]. It is usually associated with PAH and a large left-to-right shunt. On rare occasions, trauma [22] or tuberculosis has been implicated. In these cases, however, the aneurysms were peripheral as in coin lesions [19]. APA have also been seen after a Blalock-Taussig shunt or after pulmonary artery banding [6, 18].

Cystic medial necrosis with PAH may be found in the absence of the above mentioned causes [3, 8]. It may lead to a dissecting aneurysm of the PA [1, 20]. Eleven such dissecting aneurysms have been reported [16].

Since PAH is seen frequently and APA is rare, a congenital defect of the arterial wall is probably the cause of the lesion. In the absence of other signs of the disease, it has been considered as an incomplete form of Marfan's disease [3]. This was probably the case of our patient 2.

An isolated APA with cystic medial necrosis is probably rare. In our second case, the cystic medial necrosis was associated with a moderately elevated PAH.

Pulmonary valvular insufficiency is also rare. As an isolated lesion, it is the rarest of the valvular diseases (0.2%) [14]. Pulmonary valvular insufficiency associated with congenital or acquired heart disease is more common. The most common causes reported are: valve agenesis, valve dysmorphys (bicuspid and quadricuspid valves), left-to-right shunts, and idiopathic dilatation of the pulmonary artery. Among the acquired causes, one can distinguish the direct involvement of the valve (bacterial endocarditis, postoperative) and the indirect causes (mitral stenosis, cor pulmonale, PAH [7, 11].

The association of pulmonary valve insufficiency with a true aneurysm of the pulmonary artery has been very rarely reported, being only one of the last etiologies quoted [11, 25] and for some authors, it represents 8% of the cases of pulmonary valve insufficiency [5]. However, it has been more frequently (28%) quoted with idiopathic dilatation of the pulmonary artery [11]. The mechanism of the pulmonary valve insufficiency is usually a dilatation of the valvular annulus as found in our case 2. However, we found another mechanism in our case 1, namely, an inversion of the commissures, without annular dilatation, owing to the enormous dilatation of the origin of the PA. Therefore, the valves, although normal in appearance, could not possibly close in diastole. Considering the usual location of the aneurysm in the initial portion of the pulmonary trunk, it is surprising that this was not found more commonly in the literature.

Aneurysms of the pulmonary artery, once diagnosed, should be operated on because of the fatal outcome in the absence of surgery, right heart failure and rupture being the two major complications. Rupture into the pleural or pericardial cavity is sometimes encountered [3, 4] with a previous syndrome of dissection [20]. However, the evolution is unpredictable [13], and some cases of asymptomatic aneurysms of a long duration [23] have been reported. Once there are symptoms, surgery is mandatory. In our report, only in case 1 could the mild symptoms be clearly related to the presence of the aneurysm with pulmonary valve insufficiency since there were no associated defects as in case 2 (VSD, PDA).

Only a few cases of surgical repair of APA have been reported. Options for the surgical treatment of the aneurysm include resection and graft replacement [21] or simple aneurysmmorrhaphy [3, 8, 17]. The latter would seem preferable since it is simpler and, once the supposed cause of the aneurysm is eliminated, there should be no recurrence [8]. Isolated pulmonary valvular insufficiency and decreased RV function are rarely an indication for surgery. When associated with an
APA, massive pulmonary valvular insufficiency should, in our opinion, be treated since it may cause further aneurysmal dilatation if left untreated. A valvular replacement by a homograft [26] or a bioprosthesis is usually performed. We used commissural resuspension by aneurysm resection down to the annulus and aneurysmorrhaphy in the presence of normal annulus size. It is technically conceivable that annular dilatation could also be treated since a residual valvular leak, if mild, should not be detrimental.

References


Received for publication: November 17, 1986

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