Systemic to pulmonary vascular malformation

H. M. M. Pouwels*, B. K. Janevski**, O. C. K. M. Penn†, H. T. Sie‡, G. P. M. ten Velde*


ABSTRACT: A case is reported of life-threatening haemoptysis as a result of an anomalous communication between a bronchial artery and pulmonary vein, demonstrated by angiography. The patient recovered following bilobectomy of the right lower and middle lobes.

When a systemic artery is involved in an arteriovenous malformation of the lung, haemodynamics are different compared with those present in malformations fed by the pulmonary artery. This implicates other clinical features, options for surgical intervention and prognosis. In reviewing the literature, a relationship with Rendu-Osler-Weber disease is absent in these specific malformations.

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Arteriovenous malformations of the pulmonary vascular bed are anomalies characterized by vascular shunts in normally ventilated lung tissue. Several hundred cases have been described in the literature, under a variety of names, such as pulmonary angioma, haemangioma, hamartoma, arteriovenous fistula and pulmonary aneurysm.

Recently, the most correct designation was proposed by BURKE and RAFFIN [1] (1986) as pulmonary arteriovenous malformation (PAVM). This term covers a wide variety of vascular lesions, varying in size from microscopic defects to large aneurysms, single or multiple, with or without systemic arterial supply [2].

Approximately 4% of all congenital PAVMs, have a feeder artery arising from the systemic circulation, but the bronchial artery is very rarely involved [3]. PAVM with a systemic source is considered to be a specific entity, as will be discussed below. Our patient belongs to this rare group, with an anomaly of unusual and dramatic presentation. As far as we know, this is the third reported case in which the afferent blood vessel consisted of a bronchial artery.

Case history

A 21 yr old male was admitted to our hospital because of haemoptysis. In the morning, just after awakening, he coughed up a few cups of blood and became short of breath. Previously, he was in good health and had no complaints of exertional dyspnoea.

On physical examination, a pale dyspnoeic young man was seen, coughing up red blood (>600 ml). No signs of cutaneous telangiectasia, finger clubbing or cyanosis were present. No murmurs were heard on auscultation of the thorax. Initially, breath sounds were normal, except for some crackles at the right lower base. This was followed by dullness on percussion and diminished breath sounds one hour later.

The electrocardiography (ECG) showed sinus rhythm. Blood gas analysis measured an arterial oxygen saturation (Sao\(_2\)) of 71% (because of blood aspiration). The Sao\(_2\) increased to 96% with oxygen 3 l·min\(^{-1}\) by nasal tube, and the patient's condition then stabilized.

A chest radiograph revealed a sharply defined shadow in the right lower lobe and a somewhat prominent right hilus, located downwards as well as a decreased size of the lower lobe (figs 1 and 2).

Fig. 1. – The posteroanterior projection of the chest film demonstrates a sharply defined density in the right cardiophrenic angle and somewhat prominent right hilus, which is located downwards.

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On rigid bronchoscopy blood clots obstructing the right lower lobe bronchus were seen. Digital subtraction angiography of the pulmonary arteries revealed no abnormalities.

Subsequently, a selective arteriogram of the bronchial arteries was performed (fig. 3). The arterial, capillary and venous phase of the left bronchial arteriogram were normal. However, the right selective bronchial angiogram revealed a hypertrophic and tortuous right bronchial artery, with hypertrophic branches running towards a large malformation in the right lower and middle lobe. The venous drainage of this malformation occurred through the pulmonary veins from the middle and lower lobes toward the left atrium.

With this procedure a left-to-left shunt was established, forming a communication between the right bronchial artery and pulmonary vein, through a vascular mass.

One day later the patient coughed up 400 ml bright red blood and his situation became precarious. After haemodynamic parameters had been stabilized, a surgical intervention was performed.

At thoracotomy, large convolutions of expanded bronchial arteries were seen at the dorsal side of the lower branch of the right bronchus. Side branches to the upper lobe were carefully dissected, and bilobectomy of the right lower and middle lobe was performed. Histopathology showed almost complete consolidation of the lobes because of extensive haemorrhage. In the bronchial wall, bronchial arteries were enlarged with focal media atrophy.

No specific site of haemorrhage was found, as is often the case in pulmonary malformations.

Discussion

Several theories have been proposed for the origin of PAVM; however, its embryological genesis and natural course remain unclear [2, 4, 5]. Most malformations are believed to be congenital anomalies. PAVM with systemic blood supply is a rare lesion. In large series from the Mayo Clinic, only 3 out of 101 patients with PAVM derived their blood supply from a systemic feeder artery [6]. Boshier et al. [3] described an 'interesting variation of PAVM', in a large survey. In this series, in only 12 cases out of 350, the arterial blood supply derived from a systemic source (systemic-pulmonary arteriovenous malformation (S-PAVM)).

No relationship with Rendu-Osler-Weber disease has been mentioned in the literature when the afferent supply is from the systemic circulation. Dyspnoea and the classical triad of cyanosis, clubbing of the digits and polycythaemia, as a result of under-saturation in "conventional PAVM", are not present in systemic arterial to pulmonary venous malformation [7, 8], because it is a left-to-left shunt. This was already observed by Maier et al. [9] in 1948.
Shunts from systemic arteries to pulmonary vessels may originate from anomalous branches of the aorta and from bronchial [10, 11], intercostal, epicardial pericardiophrenic, lateral thoracic, oesophageal and, more frequently, internal mammary arteries [5, 12]. Accessory bronchial arteries may originate from these vessels and may contribute to the blood supply of S-PAVM. On physical examination the only sign of a S-PAVM is often a continuous systolic extracardiac murmur, which should not be misinterpreted for persistent ductus arteriosus. Because of lack of symptoms, the discovery of these malformations is often an accidental finding. S-PAVMs may be presented at any age, but most patients are young asymptomatic adults, predominantly male [13]. The right lung is involved more often than the left and, most importantly, they tend to be solitary, but may involve several lobes. S-PAVMs are subject to systemic pressure, and so tend to grow and are more apt to rupture. Due to the rarity of these lesions, there is no experience of their evolution. Theoretically, complications such as haemoptysis are more likely to occur [9].

In the clinical work-up, chest radiographs are usually abnormal, but lesions may be minimal and aspecific. A clear distinction between these malformations with a systemic artery supply and those without is possible with angiography of pulmonary arteries, as well as of selective systemic arteries [14].

Hence, because of different haemodynamic and clinical findings, PAVM, with and without systemic blood vessel supply, can be distinguished according to the criteria summarized in table 1.

Dines and co-workers [6, 10] and other authors proposed conservative surgical intervention in systemic artery to pulmonary malformations because of future enlargement of the shunt, risk of rupture and infection.

The majority of reported cases underwent a minimal surgical resection, including the entire removal of the malformation. Silicon spheres, gel-foam particles and autologous tissue are dangerous in these malformations, because of possible passage of the material into the pulmonary veins, with resultant systemic embolism [15].

Occlusion with balloon therapy is still an issue and depends on patient characteristics, local facilities and, most importantly, the necessary experience [16]. Spinal cord injury, oesophageal necrosis, bronchial ischaemia and left main-stem bronchial stenosis are reported as serious complications [16, 17].

Our patient represents the first case with life-threatening haemoptysis occurring in congenital systemic to pulmonary arteriovenous malformations (S-PAVMs), namely of a bronchial artery to pulmonary vein, which is in itself a rarity. The source of bleeding could be detected with selective angiography, which guided the definitive surgical treatment.

Table 1. - Differences between PAVM and S-PAVM

<table>
<thead>
<tr>
<th>PAVM</th>
<th>S-PAVM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female &gt; male</td>
<td>Female &lt; male</td>
</tr>
<tr>
<td>Relationship with ROW (40-65%)</td>
<td>No relationship with ROW</td>
</tr>
<tr>
<td>Classical trial of symptoms</td>
<td>Absent</td>
</tr>
<tr>
<td>Right-to-left shunt</td>
<td>Left-to-right of left-to-left shunt</td>
</tr>
<tr>
<td>Single or multiple</td>
<td>Single</td>
</tr>
<tr>
<td>Local or diffuse</td>
<td>Local</td>
</tr>
<tr>
<td>Herediatrity</td>
<td>Absent</td>
</tr>
<tr>
<td>Angiography of a. pulmonalis is mandatory</td>
<td>Angiography of a. pulmonalis and selective systematic arteries is mandatory</td>
</tr>
</tbody>
</table>

PAVM: pulmonary arteriovenous malformation; S-PAVM: systemic to pulmonary arteriovenous malformation; ROW: Rendu-Osler-Weber.

Pulmonary angiograms sometimes demonstrate an early wash-out of contrast, when the vascular shunt is towards the pulmonary artery. They fail to show a defect, when the vascular communication has its drainage to the pulmonary vein, as in our case. Obviously, for the same reason, right heart catheterization reveals no abnormality in these left-to-left shunts.

Selective angiography of systemic arteries is mandatory before any surgical or other medical intervention and should be performed as soon as possible.

References

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