Rare disease

Pulmonary arteriovenous malformation presenting as a large aneurysm

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Summary

We report the case of a 49-year-old woman who presented with breathlessness on exertion and iron deficiency anaemia. Evaluation revealed a large mass in the right lung which on further investigation was determined to be a pulmonary arteriovenous malformation (PAVM) with a large aneurysmal sac. The patient subsequently underwent a lobectomy with an uneventful recovery. The pathophysiology of PAVM and its varied presentations are also discussed.

BACKGROUND

Pulmonary arteriovenous malformations (PAVM) are rare vascular anomalies that are predominantly congenital in origin. They have a strong association with hereditary haemorrhagic telangiectasia (HHT). PAVM can produce symptoms by right-to-left shunting.

PAVM represents a rare but potentially correctable cause for exertional dyspnoea, and should be considered as a differential diagnosis in patients with minimal or no risk factors for coronary artery disease, and unexplained dyspnoea.

Treatment options include percutaneous angiographic closure and surgery. Currently most centres prefer angiographic closure with surgery reserved for complicated PAVM or patients with contrast allergy. Our patient underwent a successful lobectomy as the PAVM was complicated by large aneurysmal sac distal to a large feeding artery, having significant risk of balloon, plug or coil migration.

CASE PRESENTATION

A 49-year-old woman with no pre-existing illnesses, presented with easy fatigability and dyspnoea on exertion (New York Heart Association grade II) for the past 6 months. There were no associated symptoms of chest pain, palpitation or syncope, and no history of epistaxis or bleeding from other sites. General physical examination revealed gross conjunctival pallor. Cyanosis and clubbing were absent. A careful search for mucocutaneous telangiectasia, performed after the complete diagnosis, was also negative. A low-pitched bruit was audible over the right lower part of her chest. Systemic examination was otherwise normal.

INVESTIGATIONS

Preliminary laboratory investigations revealed severe anaemia (Hb 4.7 mg/dl) with deficient iron stores. ECG showed a normal sinus rhythm with atrial ectopics. Chest x-ray (CXR) showed a homogeneous mass in the right lower zone measuring 5.2×7.5 cm with well-defined

margins, abutting the lateral chest wall and away from the right border of heart and the diaphragm (figure 1). A dilated feeder vessel could be visualised faintly on the superior aspect (arrow). In the lateral view, the mass was seen measuring 9×9 cm, posterior to the cardiac shadow and overlying the spine (figure 2). Transthoracic echocardiography showed normal biventricular function with no structural abnormalities. However, a contrast study done with agitated saline (mixed with air) showed microbubbles in the left atrium 3–5 cardiac cycles after appearing in the right atrium, hinting at the presence of an extracardiac right-to-left shunt. Subsequently, coronary angiography revealed normal epicardial coronaries. A selective

Figure 1 Chest radiograph (posteroanterior view) revealing the large oval aneurysmal sac in the right lower zone. The dilated superior segmental artery (arrow) is faintly visualised.

Figure 2 Chest radiograph (right lateral view). The aneurysmal sac is seen like a money bag in the posterior part of the chest.

right pulmonary angiogram (figure 3) showed a large pulmonary arteriovenous fistula between the right superior segmental artery and the inferior pulmonary vein, with a large aneurysmal sac. Screening for gastrointestinal telangiectasia was negative.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses of patients presenting with dyspnoea and pulmonary nodules on a CXR include aspergillosis, nocardiosis, histoplasmosis, carcinoid lung, sarcoidosis, lung cancer, tuberculosis and lung abscess.

TREATMENT

The patient opted for surgery rather than percutaneous closure once the significance of the size of the aneurysmal sac and the risk of recurrence with percutaneous closure was explained in detail.

An elective lobectomy was scheduled after correction of anaemia with blood transfusions and haematinics. Through a posterolateral thoracotomy, a right lower lobectomy was done under general anaesthesia with a left endobronchial tube in situ. Severe lung adhesions were noted and turbulent blood flow could be seen with minimal thrill on the pulsating aneurysmal sac. The right upper and middle lobes were normal. Superior segment artery size was equal to the right main pulmonary artery (figure 4, left). Sequential arterial blood gas analyses were taken from the aneurysmal sac, inferior pulmonary vein, superior pulmonary vein and radial artery (table 1). They revealed significant desaturation with a low pO_2 in the radial artery, inferior pulmonary vein and aneurysmal sac with drastic improvement after surgery. The aneurysmal sac (figure 4, right) was thin, and no clot was found. Few areas of wall calcification were noticed. The postoperative period was uneventful with oxygen saturation of 97% on ambient air. Biopsy of excised specimen was consistent with pulmonary arteriovenous fistula with adjacent

Figure 3 Pulmonary angiogram. The right main pulmonary artery is seen with the superior, middle lobar and basilar arteries. The superior segmental artery is supplying the large aneurysmal sac. RPA, right pulmonary artery; SSA, superior segmental artery; MLA, middle lobar artery; BA, basilar artery; AS, aneurysmal sac.

pulmonary haemorrhage and hyperinflation. CXR before discharge showed full expansion of right lung.

OUTCOME AND FOLLOW-UP

Patient had an uneventful postoperative period. She is asymptomatic after 1 year of follow-up with no recurrence of lung shadows and no demonstrable right-to-left shunting on contrast echocardiography.

DISCUSSION

PAVM are resulted from direct communication between branches of a pulmonary artery and a pulmonary vein with no intervening lung tissue. They are the most common form of pulmonary vascular anomalies with an incidence of 2–3/100 000 population.¹ The earliest description of PAVM dates back to 1897 by Churton, who described a 12-year-old boy with symptomatic multiple bilateral PAVM.² PAVM are mostly congenital in nature with a very few causes of acquired malformations. They represent a developmental defect of the pulmonary vasculature. Approximately 80% of PAVMs are congenital, and of these 47–80% are associated with HHT. Only 15% of individuals with HHT have PAVM.^{3 4} Acquired causes of PAVM are rare and include long-standing hepatic cirrhosis, schistosomiasis, mitral stenosis, trauma, actinomycosis, Fanconi's syndrome and metastatic thyroid carcinoma. Common symptoms of large or multiple PAVMs include dyspnoea due to right-to-left shunting, epistaxis, haemoptysis which is occasionally massive, palpitations, chest pain or cough. Possible findings in physical examination include cyanosis, clubbing and polycythemia due to significant right-to-left shunting, bruits over the thorax overlying the malformation, mucocutaneous telangiectasia and anaemia due to bleeding from telangiectatic areas. Presence of significant anaemia in patients with PAVM suggests associated HHT.

Figure 4 Intraoperative image showing the dilated superior segmental artery (arrow) on the left. Dissected aneurysm is shown on the right.

Rare complications of PAVM include haemoptysis due to intrabronchial rupture, paradoxical embolism, cerebrovascular accidents, brain abscess, migrainous headache and endarteritis. PAVM can present at any age from infancy to the seventh decade, with the majority manifesting in the first three decades.

PAVM can be single or multiple, but are usually solitary. Most PAVM occur in the lower lobes, and more commonly in the left lower lobe. PAVM have an afferent supply from one of the branches of the pulmonary artery and rarely from the systemic circulation. Sources include the aorta, bronchial and intercostal arteries. The efferent limb usually drains into one or more branches of the pulmonary veins. Rarely, there may be anomalous drainage into the left atrium or the inferior vena cava. Anabtawi *et al*⁵ have classified PAVM into five groups based on the embryological development of lung and pulmonary vasculature. Group 1 consists of multiple small fistulas without aneurysm. Group 2 includes large peripheral arteriovenous (A-V) malformations. Group 3 comprises large centrally located A-V malformations and A-V malformations with anomalous venous drainage. Group 4 consists of large venous aneurysm with systemic communication or without a fistula whereas Group 5 includes anomalous venous drainage with fistulas. The commonly described solitary PAVM with a single or multiloculated sac was included in Group 2.

Chest radiography is a sensitive tool in the diagnosis and follow-up of patients with PAVMs. They usually

show a pulmonary nodule most commonly in the lower lobes with occasional visualisation of the feeding vessel. Contrast CT is more accurate in delineating the malformation and its connecting vessels. Contrast echocardiography via injection into a peripheral vein is very sensitive in detecting a right-to-left shunt, but cannot demonstrate the exact location of the shunt. It is a valuable tool on follow-up to demonstrate recurrence following embolisation or surgery. Radionuclide perfusion lung scan is a useful adjunct in the diagnosis and quantification of PAVM, but does not provide anatomical localisation similar to contrast echocardiography. Selective pulmonary angiography remains the gold standard for diagnosing PAVM and defining vascular architecture prior to embolisation or surgical resection. CT and MR angiography should be reserved for follow-up of cases of proven or treated PAVM.

Although there is evidence for progressive enlargement of PAVMs in patients over time,⁶ there is no consensus on the size of PAVM for subjecting them to surgery. Conventional indications for treatment have been demonstrable progressive enlargement, evidence of paradoxical embolism and symptomatic hypoxia.⁷ PAVM with sizes greater than 2 cm in diameter and afferent arteries more than 3 mm in diameter have also been recommended for surgery.⁸

Treatment options for PAVM include surgery and percutaneous embolisation therapy or embolotherapy. Embolotherapy is currently the preferred technique because of its less invasive nature and the ease of repeated procedures if lesions recur. Balloon, coil or vascular plugs occlusion of the feeder artery is done after localisation with angiography. With minimal morbidity, embolotherapy has now become the first choice in elderly patients, those with multiple comorbidities, multiple malformations in either lungs, and patients refusing surgery. Surgery is an attractive option in patients with PAVM who fail embolotherapy, those who develop serious bleeding complication during embolotherapy, have intrapleural rupture of the PAVM, or have contrast allergy and lesions not amenable to embolotherapy. Video-assisted thoracoscopy resection, segmental resection and lobectomy with an aim of lung conservation appear to be the techniques of choice in such situations.

Even though large PAVMs, with feeder vessels larger than 8 mm, have been closed successfully with

Table 1 ABG findings

Type	fiO ₂	pH	CO ₂	pO ₂	SO ₂	BE	Bicarb	Hb
Preoperative ABG (room air)	0.21	7.42	29	65.9	93	−4.3	18.7	13.3
Intraoperative (intubated)								
PA	0.6	7.33	40.8	58.6	87.5	−4.5	21.1	14.2
Radial artery	0.6	7.34	38.3	99.3	97.1	−4.6	20.5	13.8
Aneurysmal Sac	0.6	7.33	40.6	55.3	85.7	−4.3	21.3	14
Right IPV	0.6	7.34	37.7	67.1	91.6	−4.7	20.3	13.8
Right SPV	0.6	7.38	32.8	285.9	99.9	−5.1	19	14.1
Postoperative ABG	0.6	7.4	29	282.9	99.9	−5.5	17.8	14

ABG, arterial blood gas; BE, base excess; Hb, haemoglobin; IPV, inferior pulmonary vein; PA, pulmonary artery; SPV, superior pulmonary vein.

embolotherapy using a combination of coils and balloons, our patient underwent a successful lobectomy to avoid the risk of recanalisation (due to balloon or coil migration) or accessory artery growth which is commonly observed in such large PAVMs.^{9–13}

Learning points

- ▶ Pulmonary arteriovenous malformation (PAVM) is a rare but correctable cause for dyspnoea on exertion.
- ▶ PAVM can occur in isolation or in association with hereditary haemorrhagic telangiectasia.
- ▶ Large and/or symptomatic PAVM are prone to complications of right-to-left shunting and need treatment.
- ▶ Surgery is a safe therapeutic option with minimal mortality and morbidity and scores over embolotherapy in reducing the risk of recanalisation in a large PAVM.

Competing interests None.

Patient consent Obtained.

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