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Case Report

Large pulmonary arteriovenous malformation lost to follow-up with 10 years of asymptomatic interval growth: A case report ☆,☆☆,★

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ABSTRACT

Pulmonary arteriovenous malformations, previously considered a rare condition, have been increasingly identified in asymptomatic patients over the past 2 decades. Usually congenital and associated with hereditary hemorrhagic telangiectasia, these fistulae result in right-to-left shunting of blood by abnormal communication of pulmonary arteries and veins lacking capillary beds. Clinical findings of right-to-left shunting in the presence of feeding and draining vessels identified on imaging confirm the diagnosis, for which the first-line therapy is embolization. This report highlights the presentation and management of a large asymptomatic PAVM detected incidentally in a patient who was lost to follow-up for 10 years and represented with acute hypoxic respiratory failure secondary to a viral infection with an interval increase of PAVM size.

Introduction

Pulmonary arteriovenous malformations (PAVMs) are right-to-left fistulae commonly associated with hereditary hemorrhagic telangiectasia (HHT) [1,2]. They are often asymptomatic and incidentally found on imaging performed for alternative indications [3,4]. When symptomatic, patients often present with signs of right-to-left shunting including dyspnea, cyanosis, and digital clubbing, but severe cases may present with hemoptysis and hemorrhagic shock from PAVM rupture [5–7]. Treatment depends on the size and position of the lesion with endovascular embolization indicated as first-line treatment in all symptomatic PAVMs and

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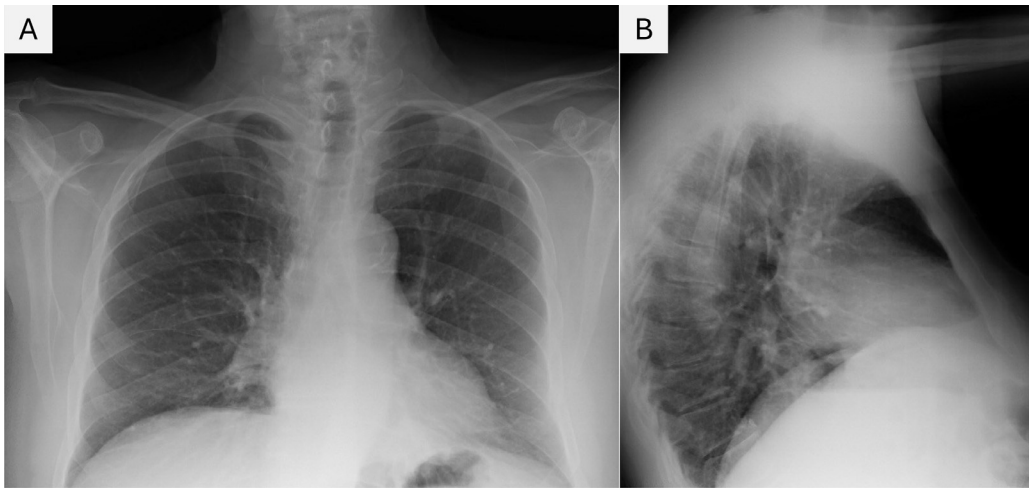

Fig. 1 – (A, B) Chest X-ray at the initial presentation, 10 years before the current episode. No abnormalities were identified in this study and the posterior cardiac contour remains uninterrupted.

for asymptomatic lesions with feeding vessels larger than 2-3 mm [3,8].

Even large PAVMs may be asymptomatic despite their increased risk of rupture, and close surveillance is recommended to monitor for the development of symptomatic disease or enlargement [1,5–8]. Here we present a case of a large, asymptomatic PAVM found incidentally in a patient without HHT who was lost to follow-up and presented 10 years later with acute respiratory failure and interval increase in lesion size.

Case presentation

Ten years before the current episode, a 73-year-old man with a past medical history of smoking, cerebrovascular accident, and hyperlipidemia presented with diffuse back pain, cough, and dehydration and was admitted for E. coli urosepsis following a prostate biopsy. A chest X-ray performed during the sepsis workup had no acute cardiopulmonary findings (Fig. 1). Non-contrast CT (NCCT) of the chest, abdomen, and pelvis performed for sepsis workup, diffuse back pain, and cough demonstrated a 5.3 cm mass-like consolidation within the medial left lower lobe (Fig. 2). Pulmonology was consulted and favored the consolidation to represent a mass or atelectasis and recommended outpatient follow-up. The patient was discharged with IV antibiotics but returned shortly after for rectal bleeding treated with colonoscopic banding of prolapsed internal hemorrhoids and subsequently underwent fiberoptic bronchoscopy inpatient while sedated. The following bronchoscopy with biopsy found no endobronchial mass or lesion and the patient was discharged.

Seven weeks after discharge, follow-up chest NCCT revealed no significant changes and recommended a PET-CT to further evaluate the mass for malignancy, which revealed no abnormally increased tracer uptake in the well-circumcised lobulated mass, favoring a benign etiology. A contrast-enhanced CT (CECT) of the chest was performed 6 months later for surveillance where the lung mass was stable in size from prior exams and demonstrated enhancement characteristics suggestive of a PAVM with a feeding artery laterally and a draining vein medially (Fig. 3). The patient was then lost to follow-up for the next decade.

The patient presented 10 years later with new-onset shortness of breath, non-productive cough, and confusion. On arrival, he was tachypneic and hypoxic with an oxygen saturation of 91%. Initial chest radiograph revealed a retrocardiac ovoid density (Fig. 4) and CECT PE protocol was negative for pulmonary embolism but demonstrated enlargement of the previously identified PAVM to 7.1 cm (Fig. 5). He tested positive for RSV and labs indicated lactic acidosis. A head NCCT did not demonstrate any masses or bleeds. Steroids, magnesium, and IV fluids were initiated to treat his acute hypoxic respiratory failure secondary to upper respiratory infection. An echocardiogram with a bubble study was performed, revealing preserved ejection fraction and no evidence of interatrial shunting.

The patient underwent left pulmonary angiography with embolization of the lower lobe arteriovenous malformation. The PAVM was accessed through the right common femoral vein, using a 5 French Sos Omni catheter (AngioDynamics, Latham, NY, USA) and a hydrophilic Glidewire (Terumo Interventional Systems, Somerset, NJ, USA). The Sos Omni catheter was then replaced over an exchange-length Amplatz wire. A 7 French by 65 cm sheath was placed in the left main pulmonary artery, subsequently, a 5 French pigtail catheter was placed in the left main pulmonary artery and selective left pulmonary arteriograms were performed. These demonstrated evidence of a large left lower lobe arteriovenous malformation with a single feeding artery (Fig. 6). The diameter of the feeding artery was at least 12 mm. Multiple oblique projections were used to identify the feeding artery. Following this, the feeding artery to the malformation was selectively catheterized using a 5 French angled catheter. Coil embolization of this vessel was then performed with multiple fibered and hydrophilic coils

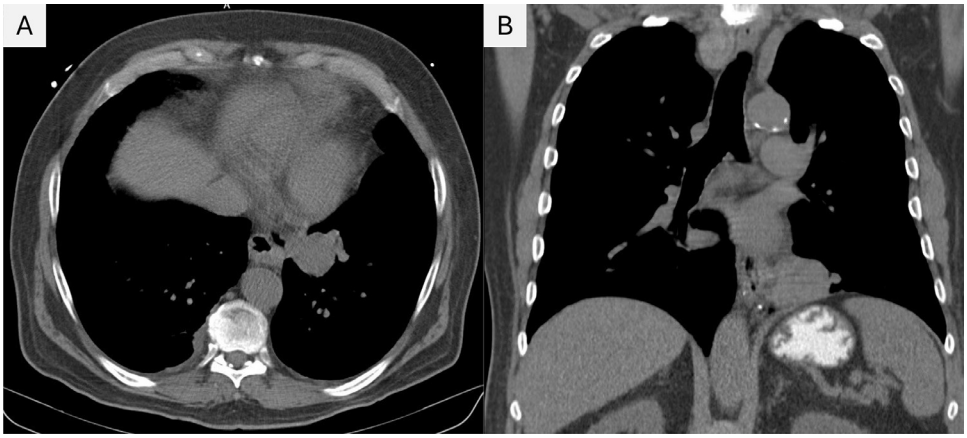

Fig. 2 – (A, B) Chest NCCT on initial presentation. The 5.3 cm retrocardiac mass abutting the diaphragm is poorly characterized in this study and initially thought to represent infection, rounded atelectasis, or malignancy.

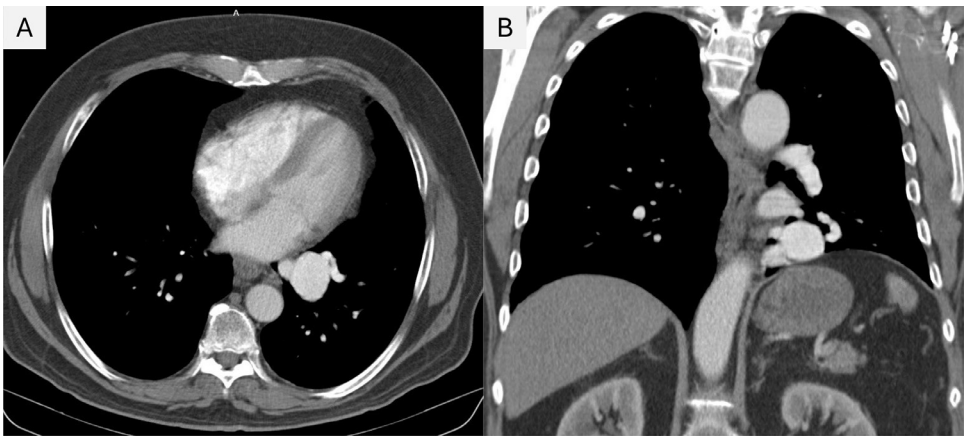

Fig. 3 – (A, B) Chest CECT Chest performed 6 months after initial presentation reveals a near-equal intensity of the thoracic mass with the blood pool suggestive of a PAVM with feeding artery laterally and draining vein medially.

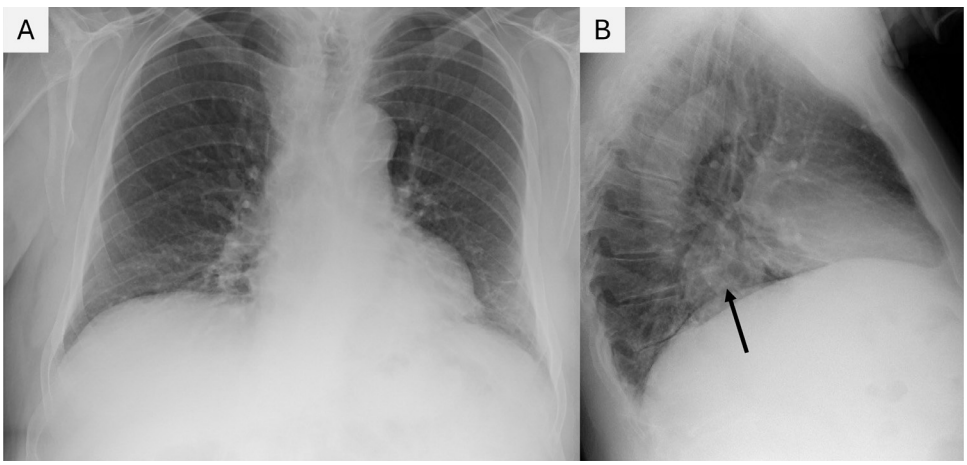

Fig. 4 – (A, B) Chest radiograph on representation 10 years after the initial episode. Note retrocardiac density is visualized only on the lateral film with obscuration of the posterior cardiac border (B, arrow).

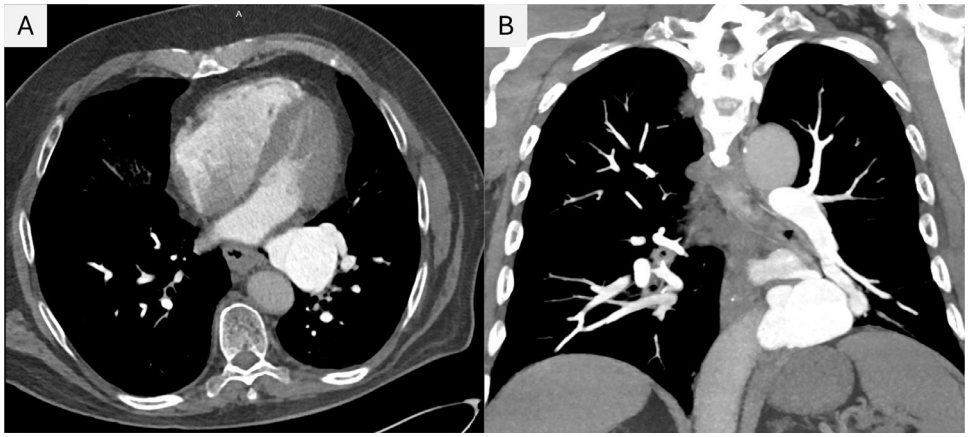

Fig. 5 – (A, B) CECT PE protocol performed on representation 10 years after the initial episode demonstrating interval PAVM enlargement now to 7.1 cm and lateral feeding vessel branching from the left pulmonary artery.

measuring in size from 10 mm to 14 mm. Post-coiling angiography demonstrated no further filling of the sac, and the coils were seen to be located appropriately.

Discussion

In the present case, the etiology of the lung mass was not identified early. This patient received NCCT initially during a workup for sepsis and back pain, resulting in poor characterization of the incidentally found mass. Standard workup for a possible chest mass typically involves CECT which would have revealed the vascular nature of this lesion. The mass was initially to be followed outpatient but when the patient returned shortly after discharge with rectal bleeding, bronchoscopy was performed with colonoscopy. NCCT performed after discharge recommended further malignancy workup with PET-CT, resulting in further delay in CECT evaluation and diagnosis of PAVM before the patient was lost to follow-up. In this patient who did not present with acute kidney injury, a history of reactions to contrast media, or other apparent contraindications for contrast-enhanced imaging, this sequence of events highlights the importance of CECT for the characterization of a chest mass and prevention of an unnecessarily delayed, invasive, and expensive workup, including in this case bronchoscopy, repeat NCCT, and PET-CT.

Pulmonary arteriovenous malformations, also known as pulmonary arteriovenous fistulae, are right-to-left shunts caused by abnormal connections between pulmonary arteries and pulmonary veins that lack capillary beds [1]. In the general population, clinically significant PAVMs are uncommon, with a prevalence of 1 per 2600 individuals found in a retrospective study of patients undergoing lung cancer screening [3]. Up to 85% of PAVM cases are associated with hereditary hemorrhagic telangiectasia, resulting from abnormal angiogenesis throughout the body [1,8]. About 30%-50% of HHT patients have PAVMs, but 10%-20% of PAVM cases lack signs of HHT or other systemic disease. In the general population, PAVMs are more common in females (1.5-1.8:1 M:F ratio), primarily unilateral (90%), and two-thirds are found in the lower lobes [9]. PAVMs in HHT are more evenly distributed across sex and lung lobes and are more often multifocal [10]. PAVMs can be classified as simple (most common), where a single segmental artery drains into one or more veins, complex, where multiple segmental arteries drain into multiple veins, or rarely, diffuse, with hundreds of malformations present.

PAVMs are congenital in more than 80% of patients, with most associated with HHT [1,11]. Rarely, they are acquired due to iatrogenic causes such as congenital heart defect repair, trauma, mitral stenosis, infections such as schistosomiasis, and hepatopulmonary syndrome, among others [9,11,12]. Right-to-left shunting results in reduced oxygenation, the severity of which is determined by the degree of shunting, and incomplete filtration of the pulmonary blood, which may result in paradoxical emboli [13].

Often patients with PAVMs are asymptomatic and found incidentally during adulthood on CT performed for another indication [3,4]. Symptomatic patients most often have dyspnea on exertion, palpitations, cough, and chest pain [10,13,14]. Patients with HHT may also present with epistaxis or hemoptysis, though this may also occur with PAVM rupture. Patients may also present with neurologic symptoms or complications such as transient ischemic attack, stroke, or brain abscess [4]. Physical exam features include cyanosis, digital clubbing, and pulmonary vascular bruit, and patients with HHT may show telangiectasias or hemangiomas [1,10,13,14].

PAVMs should be suspected in patients with a personal or family history of HHT or signs of right-to-left shunting such as digital clubbing, cyanosis, or dyspnea on exertion. Chest X-ray classically shows a round, well-circumscribed, soft tissue density that may be associated with enlarged vessels [1,8,10,14]. Noninvasive options to detect right-to-left shunting include arterial blood gases with $PaO2 < 85$ mm Hg suggesting a significant shunt (> 5%) is likely. Transthoracic contrast echocardiography with bubbles present in the left atrium is considered positive for shunting. CT with or without contrast can detect anatomic details where simple PAVMs appear as a single, well-circumscribed nodule with a single feeding artery and a single draining artery [14]. Pulmonary angiography can also confirm

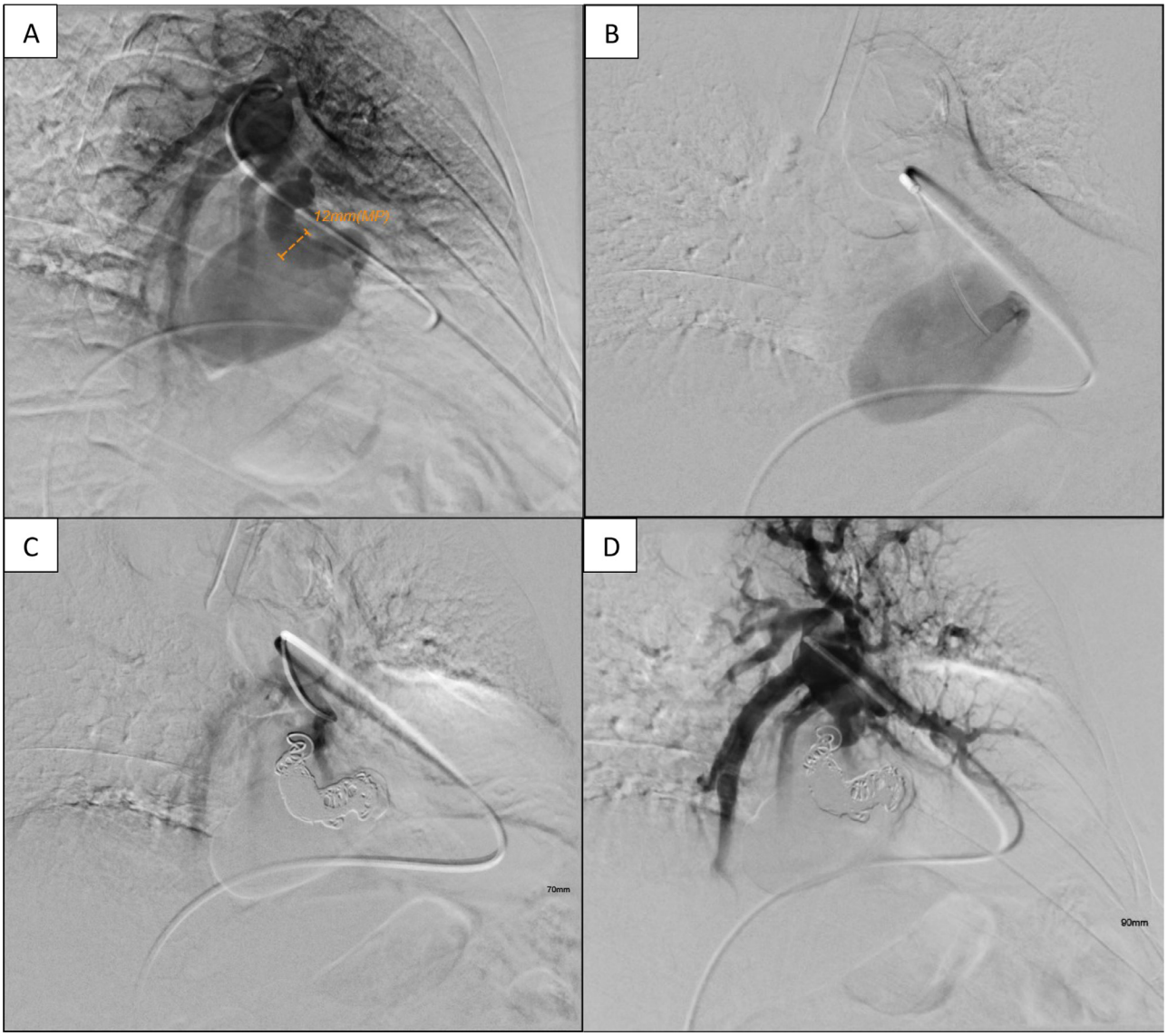

Fig. 6 – (A-D) Pulmonary angiography of the PAVM with subsequent embolization of the feeding vessel. (A) Angiography of the left main pulmonary artery with identification of feeding vessel measuring 12 mm. (B) Oblique view with selective catheterization of the feeding vessel. (C) Coil embolization of the feeding vessel. (D) Postembolization angiography of the left main pulmonary artery demonstrated no further filling of the PAVM.

the diagnosis before or concurrently with embolization. Biopsies are associated with an increased risk of severe bleeding and stroke and are not recommended in the routine workup of PAVMs [15]. Differential diagnosis includes other causes of right-to-left shunts, such as congenital heart disease or hepatopulmonary syndrome, and radiologic mimics include other causes of pulmonary aneurysmal disease, pulmonary varices, pulmonary arterial collaterals, and nonvascular abnormalities including bronchocele or tumors [11,16].

The first-line treatment for PAVMs is embolization which has a high success rate (95%-100%) and low rate of complications (most commonly pleurisy) [8,13,14]. Embolization is indicated in all symptomatic patients and asymptomatic patients with feeding vessels larger than 2-3 mm [8,9,17]. In patients who are not candidates for embolization therapy, local resection or segmentectomy may be performed. This is indicated for patients with failed embolization, serious bleeding, small feeding arteries (< 2-3mm), or allergies to contrast or embolization devices [18]. Patients who receive embolization therapy should be followed with CT imaging in 6-12 months then every 3-5 years [1,17]. Follow-up for untreated patients with asymptomatic disease can be determined by clinical judgment and it is suggested to follow the lesion for progression every 1-5 years with CT imaging.

Patient consent

Informed consent was obtained from the patient for publication of this case report and any associated images.

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