42 Pulmonary Arteriovenous Malformations: The Red and Blue Deadly Pool

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Introduction

PAVMs are rare pulmonary vascular anomalies which consists' of abnormal communications that provide a right to left shunt between the pulmonary arteries and veins without intervening capillaries. [1,2]It was recorded as 2 - 3 cases per 100000 populations, with male to female ratio ranging from 1:1.5 to 1.8. [2] Clinical presentations range from asymptomatic to severe, including hemoptysis, hemothorax, and paradoxical emboli, with epistaxis being the most prevalent symptom, followed by dyspnea, platypnea, and hemoptysis. [3,4]

Case report

A 75 - years - old lady, who was previously well presented with a three-day history of dyspnea and non-productive cough. Upon arrival to the Emergency Department, she was alert, conscious, not cyanotic, but appeared tachypneic, tachycardic with a regular heart rate and hypoxemic. Otherwise, she was normotensive and afebrile. There were no heart murmurs or pulmonary bruits detected. The other findings of cardiovascular, respiratory, abdominal, and musculoskeletal examinations were unremarkable. In the initial blood investigations, full blood count demonstrated a minimally raised total white cell with a normal hemoglobin and platelet. Her eGFR was 43 mL/min/1.73m^2. ABG revealed that the patient had type 1 respiratory failure with mixed metabolic and respiratory alkalosis. COVID-19 test was negative.
Chest X-Ray showed patchy consolidation at the left perihilar region, thus proceeded with CTPA as pulmonary embolism was included in the differentials due to hypoxia, tachycardia, and a Wells' score of 4.5. CTPA findings was suggestive of a left upper lobe arteriovenous malformation with thrombus within. Unfortunately patient's condition worsened, necessitating for intubation as well as inotropic support during the course of admission. CT brain was then performed due to poor GCS recovery and it revealed a subacute left MCA infarct. During the course of admission, she was started on anticoagulant medications and despite that she developed episodes of atrial fibrillation. She was referred to a tertiary centre for cardiothoracic surgery, where they recommended embolectomy, a less invasive technique given her age. However considering patient's advanced age and guarded prognosis, her family members opted against surgical intervention and decided for discharge against medical advice. The patient died at home on the same day.

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Declaration of conflicts

The author(s) declare(s) that there is no conflict of interest.

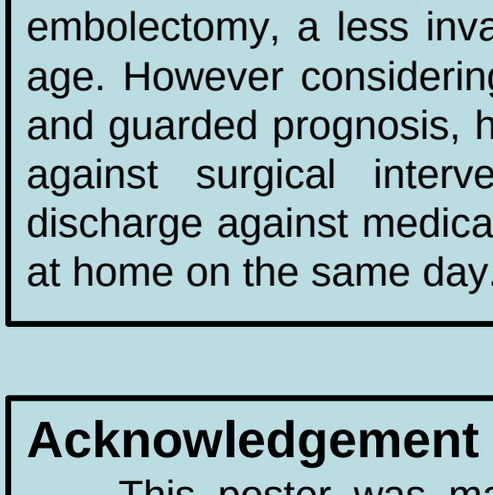

Figure 1 Gross anatomy of a large pulmonary arteriovenous malformation

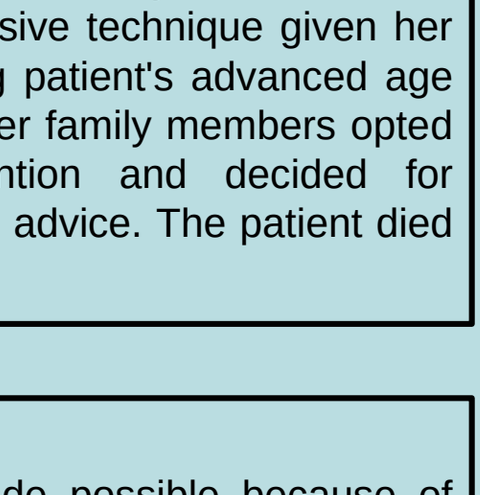

Figure 2 CTPA showed a serpentiginous mass indicating an AVM at the left upper lobe

Discussion

PAVM is linked to Hereditary Hemorrhagic Telangiectasia (HHT), also known as Osler-Weber-Rendu Syndrome[5] with 47-80% of PAVMs reported to have symptoms like epistaxis, melaena and hemoptysis.[2] It can be categorised into simple and complex based on their anatomical architectures with approximately 80% of cases are simple type which is the case seen here for this 75 year old lady whereby there is a single feeding segmental artery leading to a single pulmonary draining vein as opposed to complex types with multiple feeding arteries and receiving veins forming the malformation.[6] Solitary PAVMs are most common in the left lower lobe, followed by the right lower, left upper, right middle, and right upper lobes. PAVMs commonly affect the lower lobes of the lungs due to the increased blood flow contributed by the gravitational force. [7,8]
Common presenting symptoms of PAVM include dyspnea on exertion (27–71%), hemoptysis (2.4–20%), chest pain (1.9–17%), thrill (25–58%), cyanosis (9–73%), and clubbing (6–68%).[9] Undiagnosed patients, on the other hand, may eventually present with life-threatening consequences such as ischemic stroke, myocardial infarctions, cerebral abscesses, significant hemoptysis, and hemothorax. [7] Nonetheless, because a PAVM can be asymptomatic in many individuals, the classic trio of exertion - induced dyspnea, cyanosis, and clubbing should alert a clinician. [9,10]

Laboratory investigations of pulmonary arteriovenous malformation has shown microcytic anemia due to blood loss in patients with hereditary hemorrhagic telangiectasia (HHT) whereas liver function derangement has implied liver vascular malformation as it occurs in 32-78% of HHT patients. [11] Computed tomography (CT Chest) is a preferred diagnostic imaging in PAVM, with a higher sensitivity (90%) than Pulmonary Angiography (60%). CTPA is useful in the anatomy of PAVM, whereas contrast enhanced CT is useful in identifying vascular details of PAVM.[12]
Percutaneous embolization is the most preferred method of treatment and it can be performed as part of the investigative pulmonary angiography.[13] Diameters, the size and number of PAVMs present are factors to consider if numerous embolization attempts are required to completely occlude the multiple feeding arteries. Untreated PAVM increases the risk of morbidity and death, with thrombosis or embolism being the major cause of the following consequences. High risk complications such as brain abscesses (9%), cerebrovascular accidents (2.6–25%), hemoptysis / hemothorax (8%), and hypoxemic respiratory failure / arterial hypoxemia has been reported.[14.15]

Conclusion

PAVM is a rather uncommon clinical condition. Dyspnea, haemoptysis, epistaxis, cerebrovascular events, and hypoxemia should alert the clinician to the possibility of PAVM. Because there is a significant association between pulmonary AVM and HHT, contrast-enhanced CT Brain should be utilised to screen for cerebral AVMs or any other suspected thrombosis-related infarction.[16] CT pulmonary angiography remains the gold standard as the main diagnostic tool for PAVM. Percutaneous embolotherapy using coils or balloons is currently the most preferred way of treatment in comparison to surgical intervention such as lobectomy.[2]

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