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CASE REPORT

Pulmonary arteriovascular malformation: a rare cause of unexplained hypoxia and acute dyspnoea in young patients

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SUMMARY

Pulmonary arteriovenous malformations (PAVMs) are anomalous vascular connections between arteries and veins in the lung and comprise of two types, simple and complex. PAVMs are associated with congenital conditions such as hereditary haemorrhagic telengiectasia along with acquired causes. We present a case of a 26-year-old man who presented with dyspnoea, palpitations and decreased oxygen saturation as an initial presentation of PAVM, which was treated successively with embolisation.

BACKGROUND

Pulmonary arteriovenous malformations (PAVMs) are anomalous connections between arteries and veins in the lungs supplied by the pulmonary artery in 95% of cases.[1] PAVMs are mostly unilateral and arise from the lower lung lobes. Common clinical presentations range from cyanosis, brain abscess, stroke and fatal haemoptysis.[2] A PAVM presenting as dyspnoea is quite rare. In our case, the patient reported to ER with predominant symptoms of dyspnoea and palpitations; workup showed polycythaemia and disproportionate hypoxia. Our literature search revealed a paucity of data consistent with the clinical presentation of our case, making it imperative to report it. Our patient did not have a diagnosis of haemorrhagic telengiectasia (HHT).

CASE PRESENTATION

A 26-year-old man presented to the Emergency Department of the Aga Khan University Hospital, Karachi with symptoms of dyspnoea on mild exertion and episodic palpitations for 2 days, lasting for 10–15 min each, and recurring and resolving spontaneously. No precipitating factors or associated symptoms could be identified. The patient was a regular smoker. There was no family history of cardiac disease and no history of nasal bleed, haemoptysis, or medical or surgical history.

Physical examination revealed oxygen saturation of 85% on room air while sitting and 88% while lying down, on pulse oximetry. No clubbing was noted. A chest and precordium examination was unremarkable.

INVESTIGATIONS

A complete blood count showed polycythaemia with haemoglobin of 19.2 g/dL and haematocrit of 55%. ECG showed pre-mature ventricular contractions. A chest X-ray showed a small well-defined nodular opacity in right anterior fourth intercostal space, in the posteroanterior view (figure 1). Arterial-blood gases revealed hypoxia pH=7.44, PCO_2=22 mm Hg, PO_2=64 mm Hg with oxygen saturations=93% on 0.28% FiO_2 with an A–a gradient of 108. Echocardiography was normal. Troponins were negative. A CT of the chest with contrast was ordered to rule out pulmonary embolism or PAVM. To our surprise, the scan revealed multiple arteriovenous malformations (AVMs) in the lower lobes of the lungs bilaterally: three on the left (figure 2) and two on the right (figure 3). A nidus of AVM in right lower lobe measured 1.1 cm and the nidus of the largest AVM on left lower lobe measured 1 cm.

DIFFERENTIAL DIAGNOSIS

- Pulmonary embolism
- Valvular heart disease
- Shunts

TREATMENT

Digital subtraction angiogram was performed, which showed one major and one minor fistula on the right, and two major and one minor fistula on the left. The three major fistulas were embolised and successfully occluded with platinum detachable coils.

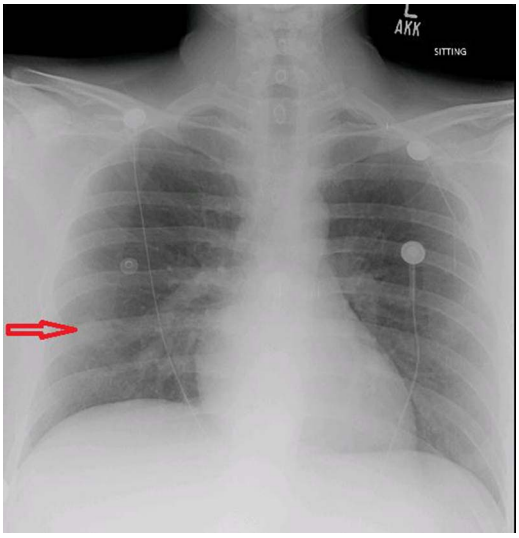

Figure 1 Chest X-ray posteroanterior view showing rounded opacity in right anterior fourth intercostal space.

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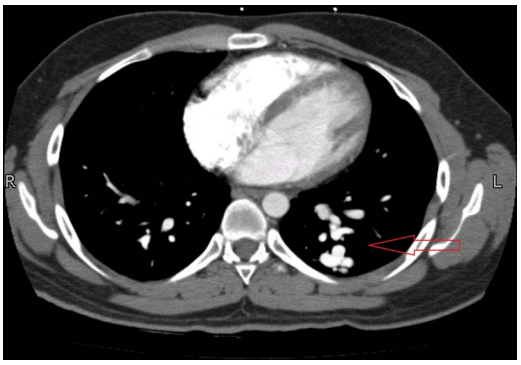

Figure 2 CT of the chest with contrast showing three arteriovenous malformations (AVMs) on the left of the lung.

OUTCOME AND FOLLOW-UP

A follow-up in the clinic showed marked improvement in the patient's presenting symptoms and maintained oxygen saturation (96%) on room air. In addition, no hypoxia or dyspnoea was observed after the patient was made to take a 6 min walk.

After the completion of treatment, follow-up in a case such as this requires evaluation of adequate oxygen delivery and signs of hypoxia. In addition, coils and feeding vessels also need to be evaluated with a repeat CT scan. Initial follow-up in uncomplicated cases is on a 3-month basis, which can subsequently be extended to 6–12 months and more until a visit to the pulmonologist is only required every 3 years unless the patient develops symptoms.[3]

DISCUSSION

PAVMs are fistulous connections between pulmonary artery and veins with high flow and low resistance. Around 70% of PAVMs are associated with HHT whereas only 20% of patients with HHT develop PAVM. Shunting of microemboli bypasses the lungs and may result in development of transient ischaemic attacks (TIAs), a cerebrovascular accident or a brain abscess.[2] The exact mechanism of the formation of PAVMs is not known, however, it is hypothesised that it is due to the failure in resorption of vascular septa.[3] A review of the Mayo Clinic experience reported a morbidity of 26–33% and mortality of 8–16% of patients when left untreated.[2]

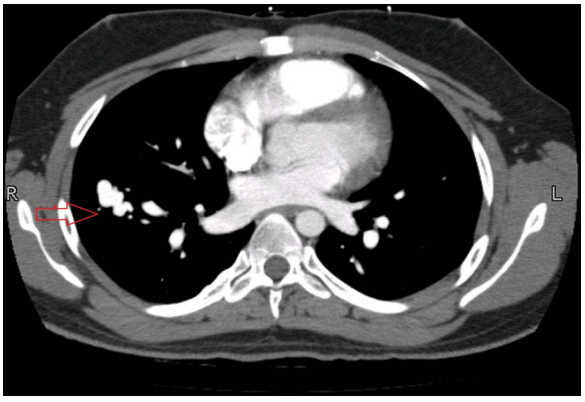

Figure 3 CT of the chest showing two arteriovenous malformations (AVMs) on the right of the lung.

Patients with PAVM lesions of >2 mm are usually well compensated and are asymptomatic despite impaired gas exchange. The slow progression of the disease allows the body's compensatory mechanisms to respond adequately: polycythaemia[4] and increased cardiac output help the body to transport adequate oxygen to the tissues.[5] Dyspnoea is a rare presentation of this disease. For patients with PAVM who present with dyspnoea, other causes should be considered. Our patient had predominant symptoms of dyspnoea and palpitation, which brought him to the hospital. He had an adequate polycythaemic response, and all other investigations for cardiopulmonary disease were negative.

Why our patient decompensated with palpitations and dyspnoea at an early age despite no comorbidities is not known, but he significantly improved postembolisation. We present this case to demonstrate that some patients with PAVMs do not cope well with the compensation required with right to left shunts, and the mechanism is still unclear.

Learning points

- Dyspnoea is a rare presentation of pulmonary arteriovenous malformations (PAVMs).
- Clinical vigilance should be maintained in a patient presenting with dyspnoea in any age group, as in this case.
- The combination of disproportionate hypoxia, polycythaemia and chest X-ray findings of nodular opacity are important indicators of PAVMs.
- In cases where haemoptysis, dyspnoea or hypoxia are unexplained, PAVM should be listed as a differential and diagnostic tests for it should be undergone.

Contributors NI wrote the manuscript and review the literature. KAR made the literature search and helped in writing the manuscript. JAK supervised and reviewed the manuscript. TUH performed the intervention on the patient.

Competing interests None.

Patient consent Obtained.

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