Case Report

Hemoptysis: Beyond routine chest computed tomography and bronchoscopy

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ABSTRACT

Hemoptysis is considered as a medical emergency which requires urgent stabilization with identification and correction of underlying etiology. Diagnosis of the cause of hemoptysis is not always readily identified after bronchoscopy and conventional computed tomography (CT) chest. Arteriovenous malformation (AVM) is a rare but important cause of massive hemoptysis which can be easily picked up by the use of double turn contrast CT chest. We here report a rare congenital AVM anomaly called Klippel-Trenaunay-Parks-Weber syndrome as a cause of massive hemoptysis and utility of double turn CT in diagnosing AVM as a cause of hemoptysis.

KEY WORDS: Arteriovenous malformations, Klippel-Trenaunay-Parkes-Weber syndrome, Klippel-Trenaunay-Weber syndrome, Parkes-Weber syndrome

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INTRODUCTION

Klippel-Trenaunay-Weber syndrome (KTWS) was first reported by Klippel and Trenaunay in 1900 as a rare congenital anomaly characterized by the triad of port-wine stain, varicose veins, and asymmetrical bony and soft tissue hypertrophy of the extremities.[1] The main abnormality was vascular malformations not only limited to skin, mucus membranes, and subcutaneous tissues but also extending to visceral organs, for example, gut, lungs, and bladder, hematuria, hematochezia, hemoptysis, and spontaneous internal bleeding have been reported in this syndrome.[2] However, the presence of arteriovenous malformation (AVM) has been named as Parkes-Weber syndrome or KTWS. Making this distinction has a significant role in management due to increased morbidity associated with AVMs.[3] Hemoptysis can be caused by angiomatous malformations in tracheobronchial tree, pulmonary thromboembolism (PTE) secondary to deep vein thrombosis (DVT) in varicose and abnormal deep veins or from AVMs in lungs.[4] Routine computed tomography (CT) scans and bronchoscopy can be normal and require CT-pulmonary angiography as well as CT aortograms to pick up the cause in such cases. We present one such rare case where double turn angiography of posteroanterior (PA) and aorta led to the recognition of this syndrome. Hence, particle embolization was done, saving the patient from unwarranted lobectomy.

CASE REPORT

A 64-year-old, male, nonsmoker, nonalcoholic with a history of hypertension on irregular treatment for 5 years, presented to the emergency room with a history of a cough, moderate hemoptysis, and shortness of breath for the past 3 days. There was no history of fever, chest pain, or recent chest trauma or thoracic surgery. In the past, the patient had recurrent episodes of mild to moderate hemoptysis since last 8 years which used to resolve after medications

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from local physicians. There was a history of massive hemoptysis 2 years ago requiring admission in intensive care unit for 3 days. Chest X-ray (CXR) was normal, and high-resolution CT-thorax was done which reported ground glass opacity in the right lower lobe followed by fiber-optic bronchoscopy which showed fresh bleeding and a blood clot in the right lower lobe bronchus [Figure 1a-d]. Bronchoalveolar lavage for cytology and 6 weeks culture for tuberculosis was negative. Subsequently, the patient had three more episodes of self-limiting hemoptysis which were managed conservatively.

On admission, his vitals were stable except tachycardia. General physical examination revealed markedly dilated, and tortuous veins in left lower limb with soft tissue thickening confirmed on X-ray of left leg [Figure 2a-c]. Routine blood examination showed normal leukocyte count with shift to the left. Chest radiograph was essentially normal. The patient was managed with intravenous fluids, tranexamic acid, cough suppressants, and antibiotics. Bilateral lower limbs venous Doppler revealed multiple tortuous vessels in posterior and anterolateral aspects of left leg with no evidence of superficial thrombophlebitis or thrombus. CT-pulmonary angiography showed no evidence of thromboembolism, but CT-aortic angiography showed an aberrant arterial vessel arising from the right lateral wall of descending thoracic aorta supplying pleura and parenchyma of superior segment of the right lower lobe with an AVM [Figure 3a-d]. Contrast echocardiography with agitated saline was negative for intrapulmonary shunting. The patient underwent right-sided bronchial artery embolization, and two collaterals from aorta were successfully embolized using polyvinyl alcohol particles with complete success [Figure 4a and b]. Postprocedural period was uneventful. The patient was advised compression stockings for the management of varicose veins. On follow-up visit, the patient had right-sided pleuritic chest pain with normal CXR which required nonsteroidal anti-inflammatory drugs (NSAIDs). There were no further episodes of hemoptysis and patient has remained asymptomatic for the past 1 year.

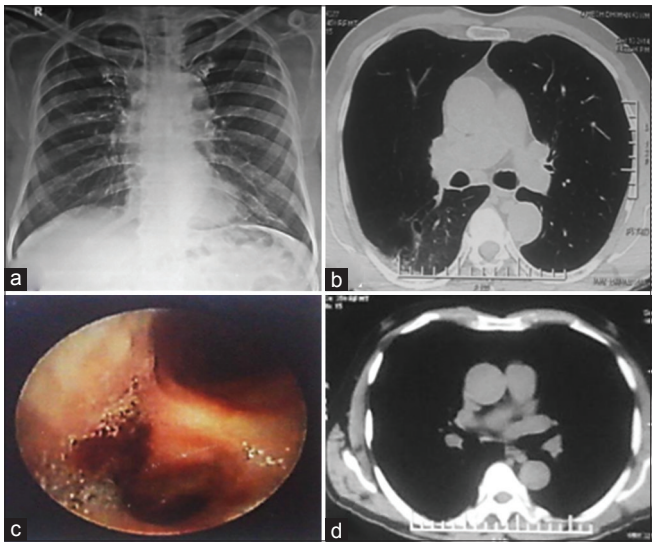

Figure 1: (a) Parenchymal abnormality on X-ray chest, (b and d) ground glass opacity in the right lower zone which is not seen in mediastinal window, (c) fiber-optic bronchoscopy showing active bleeding and clot at the right lower lobe bronchus

DISCUSSION

Moderate to massive hemoptysis is a respiratory emergency as it carries significant mortality and hence requires definite intervention to improve outcomes. Although malignancy, infections (tuberculosis or fungal infections) and bronchiectasis remain the most important and frequent causes of hemoptysis readily diagnosed by chest CT scans and bronchoscopy, there are times when even these tests are noncontributory and unable to clinch the diagnosis. Recurring hemoptysis in such situations is frustrating and leads to repeated investigations increasing patient's anxiety and cost. Pulmonary AVMs (PAVMs) though rare, are under suspicion in these cases and requires digital subtraction angiography which is frequently not done due to low suspicion of index and nonavailability at most of the centers.

PAVM has been described classically as an entity with triad of hemoptysis, cyanosis, and clubbing resulting from dysregulated angiogenesis seen in genetically determined individuals but the role of genetics in isolated PAVMs is unknown.[5] The three essential structural elements of PAVMs are the arterial supply ("feeder vessel"), a draining vein and the intervening aneurysmal sac involving pulmonary circulation. They are mostly asymptomatic but may present with hemoptysis in 4%–18% cases. Respiratory failure and pulmonary hypertension are complications infrequently seen.[6] Routine CXR may reveal nodules but may be normal if PAVM is <2 cm in size. Contrast echocardiography using agitated saline showing a delay in the appearance of the bubbles beyond 4–5 cardiac cycles into left chambers of heart confirms intrapulmonary shunting and is used as screening tool for PAVMs.[7] However, it may be falsely negative if feeding vessel is from systemic circulation as was in our case. However, clinicians should be aware of this rare syndrome where the physical signs are so striking that diagnosis should rarely be missed.

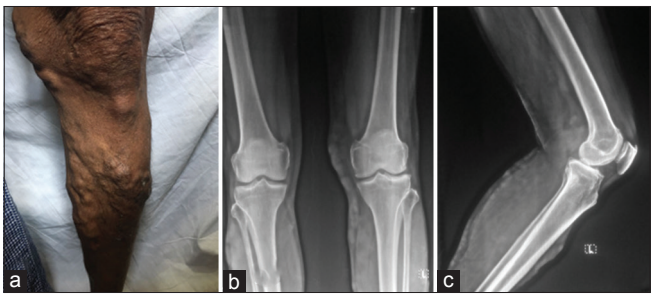

Figure 2: (a) Markedly dilated and tortuous veins in the left lower limb with soft tissue thickening (b and c) X-ray lower limb AP and lateral view showing soft tissue hypertrophy with no bony changes

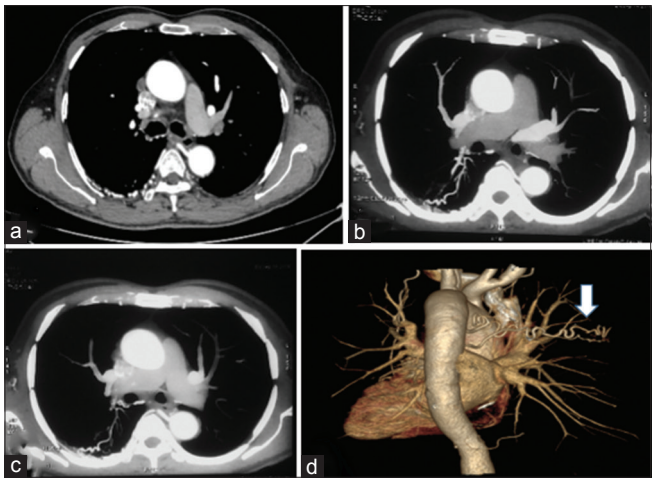

Figure 3: (a-c) Computed tomography-aortic angiography showed aberrant arterial vessels arising from right lateral wall of descending thoracic aorta supplying pleura and parenchyma of superior segment of the right lower lobe with an arteriovenous malformation, (d) computed tomography reconstruction image showing the dilated tortuous vessels arising from the descending thoracic aorta

CT chest angiograms are recommended, but again if supply to PAVMs is from systemic artery, it may be missed in PA angiography and needs double turn angiogram recordings, which includes complete aortogram along with PA angiogram. In our case, we could demonstrate PAVM from aberrant systemic artery from descending thoracic aorta. However, the possibility of sequestration could not be ruled out completely even with no evidence of pulmonary parenchymal abnormality, had there not been "telltale" signs of KTWS.

Currently, conflicting opinion exists whether to separately designate the original triad of "nevus vasculosus osteohypertrophicus" as Klippel-Trenaunay syndrome (KTS) (needs 2 out of three criteria to be met) and the triad with the addition of AVM as Parkes-Weber syndrome. All three signs of K-T syndrome: port-wine stain, varicose veins, and bony and soft tissue hypertrophies are seen in 63% of patients while 37% have 2 out of 3 features.[8] Our patient had soft tissue hypertrophy of lower limb with varicose veins confirming KTS and demonstration of PAVM in the right lower lobe fulfilling criteria of Parkes-Weber syndrome. It generally affects single limb and lower extremity being most frequently involved as in our case. Magnetic resonance imaging studies are helpful in documenting measurements of extremities and progressive growth of bones and soft tissues.[9] Although the association between the angiogenic factor gene *AGGF1* and KTS appears to be significant cases are sporadic, and our patient also had no history of such affliction in any family member.

Treatment for KTWS is conservative and symptomatic. A compression garment or intermittent pneumatic compression pumps are recommended for varicose veins as they also protect the limb from trauma and prevention of DVT which is a well-recognized complication which may lead to PTE. Surgical intervention in the treatment of varicosities and venous malformations is controversial and endovascular lasers are being increasingly used.[10]

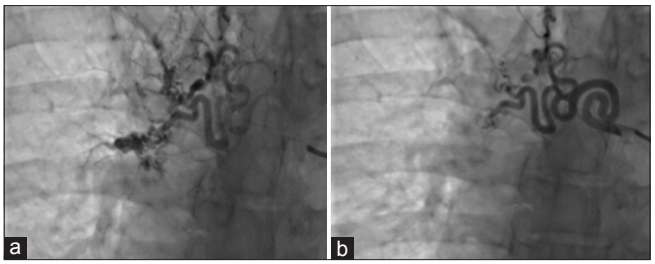

Figure 4: (a) Dilated and tortuous right-sided bronchial artery collaterals from the descending aorta during angiogram, (b) which was successfully embolized with polyvinyl alcohol particles

PAVMs may cause massive hemoptysis in 2% cases and has been traditionally treated with lung resections which led to considerable morbidity and mortality with the loss of viable lung tissue as well as chances of reoccurrences in residual lung. Coil embolization was introduced by Terry *et al*. and soon achieved a success rate of 93% and is now considered a procedure of choice in the management of PAVMs provided feeding vessel is >3 mm in size. Embolotherapy is being done now with vascular plugs and PVA (Polyvinyl Alcohol) particles which were used in our case. Reported revascularization rate of 15% require follow-up of these patients and may necessitate re-embolization. Postembolotherapy, complications such as air embolism, perforation of PAVM, cerebral infarction, pleurisy, pleural effusions, are reported[11-15] and our patient had pleuritic chest pain 1 week later but subsided with NSAIDs and repeat CXR was clear.

In our case, right lower lobe lobectomy was contemplated with laser resection of varicose veins, however, realizing the diagnosis of KTWS, the complete management changed with conservative approach for varicose veins and interventional embolotherapy for PAVMs.

CONCLUSION

This case brings an important message to the clinicians that every case of moderate to massive hemoptysis should be evaluated thoroughly, and when chest CT scans give no clues to the origin and cause of bleeding into the lungs, double turn CT angiograms should be done to rule out PAVMs.

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Conflicts of interest
There are no conflicts of interest.

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