

Hemothorax secondary to ruptured pulmonary arteriovenous malformation: A combined stitch in time saves nine

Sir,

Spontaneous hemothorax due to ruptured pulmonary arteriovenous malformation (PAVM) is uncommon. Tube thoracostomy may be delayed until a definitive procedure like embolization is performed. Embolization of PAVM in experienced hands is the definitive management. We report a case of spontaneous right-sided hemothorax secondary to rupture of subpleural PAVM that was managed with coil embolization followed by thoracoscopic-assisted removal of clots, resulting in early recovery.

A 28-year-old, previously healthy male presented to emergency with severe right-sided chest pain and breathlessness. He denied any history of trauma. He gave a history of recurrent spontaneous nose bleeds since childhood. On examination, he was tachypneic and pale with right-sided tracheal shift and generalized stony dullness over right hemithorax with decreased breath sounds. This was confirmed on a chest radiograph [Figure 1a]. Blood investigations showed hemoglobin of 7.7 g/dl with hematocrit of 25%. Other blood investigations were normal. Oxygen hemoglobin saturation on room air was 86%. Diagnostic thoracentesis was done which revealed hemothorax.

Emergency computed tomography pulmonary angiography (CTPA) was done for further valuation. Noncontrast CT [Figure 2a] revealed large right hemothorax with

collapse of entire right lung. CTPA [Figure 2b] revealed a contrast filled sac in the upper part of collapsed right lung, which was supplied by a branch of superior division of the right pulmonary artery.

Right-sided tube thoracostomy was deferred until embolization so as to prevent loss of tamponade effect of hemothorax over the AVM.

Digital subtraction angiography [Figure 3a] confirmed the presence of the arteriovenous malformation with no active extravasation. The feeding artery superselectively cannulated and embolized using interlocking microcoils of size 6 mm × 15 mm and 4 mm × 12 mm (0.018" diameter, Boston Scientific, USA) [Figure 3b]. Care was taken to do the embolization using anchoring technique in which initial few centimeters of coil is anchored into a small arterial branch with rest of the coil placed into main lumen of feeding artery.

With gradual drainage of hemothorax, dyspnea improved with decrease in oxygen requirement. Repeat chest X-ray showed partial expansion of the right lung with persistent opacification of mid and lower right hemithorax.

On 7th day of post presentation, video-assisted thoracoscopic (VATS) guided evacuation of large amount of organized clots done from pleural cavity along with decortication of pleura. Post-VATS chest drain was removed on day 8 and chest X-ray [Figure 1b] was done which revealed

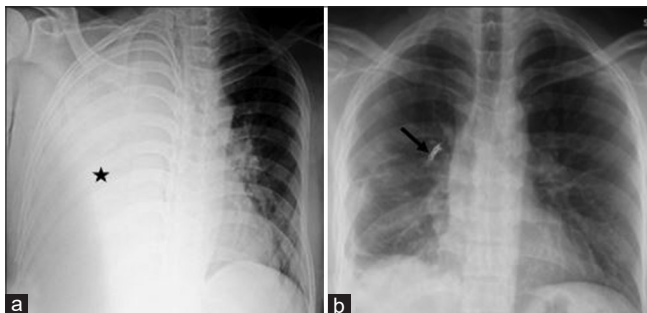


Figure 1: Chest radiographs. (a) At the time of presentation, complete opacification of right hemithorax (asterisk) with left side mediastinal shift. (b) At the time of discharge showing coil mass (black arrow) with near-complete lung expansion and mild residual pleural thickening in right costophrenic angle

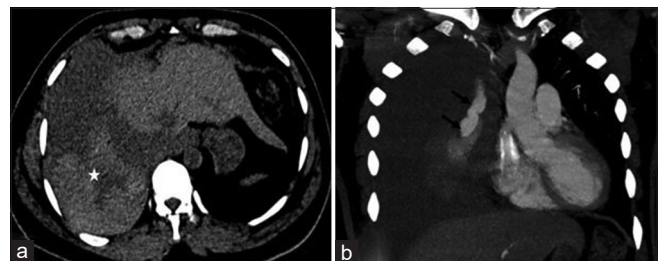


Figure 2: Computed tomography pulmonary angiography: Noncontrast axial (a) image reveal fluid in right pleural space with internal hyperdensity (asterisk) suggestive of hemothorax. (b) Computed tomography angiography coronal view shows contrast filled outpouching (black arrow) suggestive of pulmonary arteriovenous malformation in collapsed right upper lobe



Figure 3: Digital subtraction angiography. (a) Selective angiogram of the right pulmonary artery showing arteriovenous malformation arising from right segmental pulmonary artery with venous sac (black arrow). (b) Postembolization angiogram showing no opacification of arteriovenous malformation with coil mass (white arrow) and retained contrast in venous sac (double arrow)

complete expansion of the right lung with minimal residual pleural thickening. Screening for cerebral AVM with MRI brain was negative. The patient was discharged on day 12 in satisfactory condition. The patient was on regular follow-up for 6 months with no similar or new symptoms and no recanalization of AVM on follow-up CT scan.

PAVMs are structurally abnormal vessels that provide direct capillary-free communication between the pulmonary and systemic circulations and hence an anatomic “right-to-left” shunt.^[1]

About 90% of individuals with PAVMs have hereditary hemorrhagic telangiectasia (HHT).^[2,3] HHT is inherited as an autosomal dominant trait and most commonly results from a pathogenic sequence variant in ENG-encoding endoglin (HHT type 1/HHT1), ACVRL1-encoding ALK-1 (HHT type 2/HHT2), or SMAD4.^[4] Non-HHT-related PAVMs are most commonly sporadic, or secondary to hepatopulmonary syndrome, caval pulmonary shunts, or trauma.^[2,3]

PAVM can present with dyspnea, asymptomatic hypoxemia, chest pain, and hemoptysis.^[1,4] Paradoxical embolism from right-to-left shunt may lead to cerebral abscess, stroke, and rarely myocardial infarction.^[5] Spontaneous hemothorax is a rare complication of PAVM.^[6]

Rupture of PAVM leading to hemothorax is rare and is usually seen in pregnancy and when PAVMs are perfused at systemic pressures by bronchial artery or nonbronchial systemic arteries usually after embolization.^[1,6] The tamponade provided by the hemothorax may prevent exsanguination; hence, drainage of blood from the pleural space should be delayed until the PAVM has been treated.^[7]

Management of PAVM includes endovascular embolization and surgery. Embolization has become the preferred method of treatment for symptomatic PAVMs and all radiologically visible PAVMs with not necessarily diameter >3 mm.^[3] Surgery is reserved for cases in whom embolization is not feasible.^[2]

Endovascular management involves selective embolization of feeding artery supplying the AVM. Embolization can be done using vascular plug or metallic coils or a combination of both.

The conventional initial management of hemothorax is tube thoracostomy. Based on the volume of initial output, the next step may be either observation or urgent thoracotomy. In our case, VATS was done to remove residual clots which prevented the occurrence of fibrothorax and empyema.

Our case highlights the fact that PAVM can present with massive spontaneous hemothorax. Tube thoracostomy may be deferred until a definitive procedure like embolization is performed. Embolization of PAVM in experienced hands is a minimally invasive method to treat it. Early VATS after embolization leads to rapid recovery with complete expansion following hemothorax.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Submitted: 13-Jul-2019 Accepted: 09-Sep-2019

Published: 04-May-2020

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DOI:

10.4103/lungindia.lungindia_321_19

How to cite this article: Khera PS, Chauhan N, Garg PK, Choudhary R, Deokar K, Niwas R. Hemothorax secondary to ruptured pulmonary arteriovenous malformation: A combined stitch in time saves nine. *Lung India* 2020;37:272-4.

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