Pulmonary arteriovenous malformation: An unusual cause of hemothorax

Sir,

A non-smoker 38-year-old male presented with complain of sudden onset of progressive chest pain and breathlessness of 12-hour duration. Chest pain was sharp and localized to right anterior chest region. Patient denied any history of trauma sustained, fever, hemoptysis, expectoration, orthopnea, sweating, vomiting, abdominal pain and/or epistaxis. His personal, family, occupational and drug history were non-contributory.

On presentation to us, patient appeared in respiratory distress, his pulse was 108 beats/minute, blood pressure 110/80 mmHg and respiratory rate of 26 breaths/minute. His resting SpO_2 was 93% on room air. Clubbing was noted in all the fingers but cyanosis was absent. There was no pedal edema. Skin and mucus membrane examination were also normal. Examination of chest was suggestive of massive right-sided pleural effusion. Cardiovascular, abdominal and central nervous system examination were unremarkable.

Complete blood counts, routine urine examination, renal and hepatic functions including coagulation profile were within normal limits. Hemoglobin was 10.6 gm%. Electrocardiogram showed sinus tachycardia. Skiagram chest showed opaque right hemithorax with shift of mediastinum to left side. Left lung field was apparently normal. Diagnostic thoracocentesis revealed grossly hemorrhagic aspirate with hematocrit of 24% and proteins 5.2 gm%. Differential counts of pleural fluid showed polymorphs 64% and lymphocytes 36%. Pleural fluid cytology was negative for malignant cells.

In view of the presence of clubbing with hemothorax, closed pleural biopsy was performed with Abrams needle with aspiration of remaining pleural fluid. Histopathological examination of pleural biopsy was not conclusive. Follow-up chest X-ray after pleural biopsy showed minimal pleural effusion.

Contrast-enhanced computed tomography (CECT) of thorax was performed to know the state of underlying pleural space and lung parenchyma. This study showed residual small pleural effusion on right side with conglomerate of multiple soft tissue masses in peripheral part of right anterior chest in relation to anterior segment of right lower lobe [Figure 1]. Contrast echocardiography with injection of agitated saline (to create bubble) in peripheral vein was done. Bubbles appeared in left atrium after fourth cardiac cycle suggestive of a right to left shunt.

CECT and echocardiography findings favoured a strong possibility of pulmonary arterio venous malformation (PAVM). Therefore, multidetector computed tomography (MDCT) thorax angiography was performed to confirm PAVM and to delineate the number and size of the feeders. MDCT angiography showed a lobulated vascular mass of 25.7 mm × 13.8 mm in right anterior lower chest with a single feeder vessel of 5.5-mm diameter. Efferent was seen draining right inferior pulmonary vein [Figures 2 and 3]. In view of a single PAVM with a feeder vessel of 5.5 mm in diameter, patient was referred to thoracic surgeon. Video-assisted thoracoscopic wedge resection of malformation was done with ligation of the feeder. Postoperatively patient had favorable recovery with normalization of his resting SpO_2 and was doing well till the fourth week of follow-up.

Spontaneous hemothorax is a rare entity. The most common cause of this condition is metastatic pleural disease.[1] Other rare causes include anticoagulation therapy, rupture of aortic aneurysm, pleural endometriosis, extramedullary hematopoiesis, hemophilia, thrombocytopenia, spontaneous pneumothorax, acute pancreatitis, and PAVM.[2]

Dyspnea, hemoptysis, cyanosis and clubbing constitute the usual presenting features of PAVM. Occasionally,

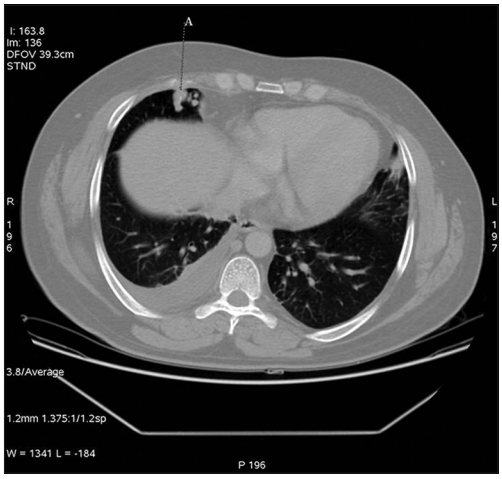

Figure 1: CECT thorax (lung window) section at just above the diaphragm showing conglomerate soft tissue nodular lesions, one with nodule of calcification in right anterobasal lung. A small amount of right pleural effusion is also seen

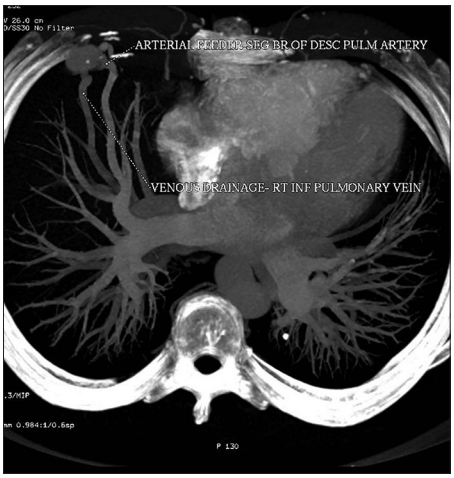

Figure 2: MDCT angiography MIP image showing a single feeder of 5.5 mm in diameter, originating from right pulmonary artery and lobulated enhancing mass of 23.7 × 13.8 mm in relation to anterior segment of right lower lobe. A single venous drainage is seen to right inferior pulmonary vein

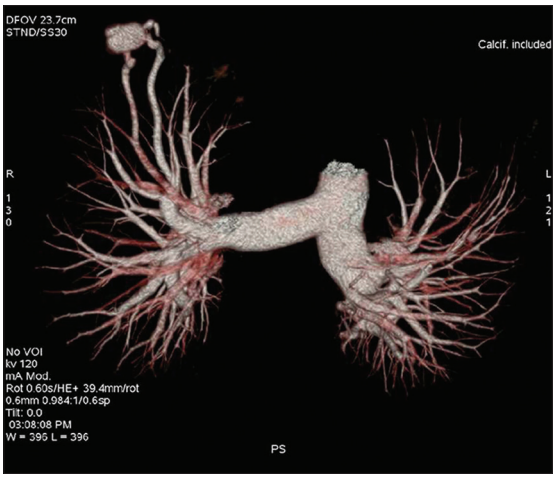

Figure 3: MDCT angiography volume rendered image showing a lobulated vascular malformation with a single feeding vessel originating from pulmonary artery and a single draining vein

patient may also present with neurological complications for e.g. stroke, transient ischemic attacks, migraine and headache. Hemothorax as a presenting feature of PAVM is a very rare occurrence. Ali *et al*.[2] found only 32 reported cases of hemothorax associated with PAVM till 2008. Half of these cases had documented HHT. On the basis of negative family history, and absent signs/symptoms like epistaxis, mucocutaneous telangiectasia HHT can be ruled out in the present case adding to its rarity.

Cottin *et al*.[3] described four patients of hemothorax in a series of 126 patients with PAVM. Ference *et al*.[4] reviewed the incidence of pulmonary hemorrhage in patients with HHT and PAVM. Six out of 143 patients had hemothorax secondary to rupture of PAVM and in all these patients hemothorax was the presenting illness. Recently, Quinones *et al*.[5] described a 39-year-old female with HHT and PAVM having hemothorax as a presenting symptom.

Presence of clubbing and hemothorax in our patient led us to initially consider the possibility of malignant pleural effusion and thence to pleural biopsy. However, pleural biopsy turned out to be negative for malignancy.

There was no other clue on clinico-radiological examination that could guide us about the possible cause of hemothorax in this patient. Therefore, we decided to have CT thorax enhanced to delineate the morphology of underlying lung and pleura. This investigation brought us closer to the etiological diagnosis of hemothorax.

Conventional pulmonary angiography and MDCT thoracic angiography were the two options before us to confirm the diagnosis of PAVM in this patient. Pulmonary angiography and MDCT angiography chest both have their own merits and demerits. MDCT thoracic angiography was preferred as it is superior in demonstrating small arteriovenous malformations, non-invasive and relatively inexpensive than conventional pulmonary angiography. Further, MDCT thorax also gives information about other pathologies in thorax, if any. But, vascular lesions may also give false positive result on this modality.[6] Contrast-enhanced MR angiography has also been found to be more sensitive than pulmonary angiography in detecting PAVM. Further, although contrast enhanced MR angiography has the advantage of acquiring MIP reconstructions of complex PAVM in multiple planes but it is inferior in delineating lung parenchymal pathology as compared to MDCT angiography.[7]

Perfusion scan and contrast 2D echo may also detect a right to left shunt but these modalities also give positive results in presence of intracardiac right to left shunts.[8] The two available options to treat this patient were embolization versus surgical excision. Risk of future recurrence and the presence of a single malformation tilted the balance in favor of surgical excision of the lesion. Video-assisted thoracoscopic wedge resection was performed as it is associated with less morbidity and mortality than conventional thoracotomy.

Manohar Lal Gupta, Mridul Gupta[1], Dinesh Kumar Vijay[2], Rajeev Bagarhatta[3]

Departments of Chest Diseases and Tuberculosis and [3]Cardiology, Sawai Man Singh Medical College, Jaipur, Rajasthan, [1]Department of Medicine, Rajarshee Chhatrapati Shahu Maharaj Government Medical College, Kolhapur, Maharashtra, [2]Department of Pulmonary Medicine, Santokba Durlabhji Memorial Hospital, Jaipur, Rajasthan, India
E-mail: drmanohargupta@gmail.com

REFERENCES

1. Mayse ML. Non malignant pleural effusion. In: Fishman AP, editor. Fishman's Pulmonary Diseases and Disorders. 4th ed. New York: McGraw Hill; 2008. p. 1487-504.
2. Ali HA, Lippmann M, Mundathaje U, Khaleeq G. Spontaneous

hemothorax: A comprehensive review. Chest 2008;134:1056-65.
3. Cottin V, Chinet T, Lavolé A, Corre R, Marchand E, Reynaud-Gaubert M, *et al*.; Groupe d'Etudes et de Recherche sur les Maladies "Orphelines" Pulmonaires (GERM"O"P). Pulmonary arteriovenous malformations in hereditary hemorrhagic telangiectasia: A series of 126 patients. Medicine (Baltimore) 2007;86:1-17.
4. Ference BA, Shannon TM, White RI Jr, Zawin M, Burdge CM. Life threatening pulmonary hemorrhage with pulmonary arteriovenous malformations and hereditary hemorrhagic telangiectasia. Chest 1994;106:1387-90.
5. Parrilla Quinones F, Marmorato Rivera R, Calderon RE, Vargas Otero P, Hidalgo Rios A. Spontaneous hemothorax: First manifestation of pulmonary arteriovenous malformation in a patient with hereditary hemorrhagic telangiectasia. Bol Asoc Med *P* R 2011;103:41-4.
6. Maldonado JA, Henry T, Gutiérrez FR. Congenital thoracic vascular anomalies. Radiol Clin North Am 2010;48:85-115.
7. Schneider G, Uder M, Koehler M, Kirchin MA, Massmann A, Buecker A, *et al*. MR angiography for detection of pulmonary arteriovenous malformations in patients with hereditary hemorrhagic telangiectasia. AJR Am J Roentgenol 2008;190:892-901.
8. Iqbal M, Rossoff LJ, Steinberg HN, Marzouk KA, Siegel DN. Pulmonary arteriovenous malformations: A clinical review. Postgrad Med J 2000;76:390-4.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.168116