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A CASE OF PULMONARY ARTERIOVENOUS MALFORMATIONS

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Abstract:

Spontaneous hemothorax is a rare entity and most commonly caused by metastatic pleural disease. Other rare causes include anticoagulation therapy, rupture of aortic aneurysm, pleural endometriosis, extramedullary hematopoiesis, hemophilia, thrombocytopenia, spontaneous pneumothorax, acute pancreatitis, and Pulmonary Arteriovenous Malformation [PAVM]. PAVMs are rare abnormalities of pulmonary vascular system characterized by an abnormal communication between the pulmonary artery and vein, resulting in a low resistant right-to-left shunt. The PAVMs have an impact on world with just 2-3 cases per 1,00,000 population. PAVMs can be either congenital or acquired and are most commonly associated with hereditary haemorrhagic telangiectasia. Approximately only 10% of cases are diagnosed in childhood, many remain undiagnosed till adulthood and are diagnosed only in 5th-6th decades of life.

Keywords:
Pulmonary Arteriovenous Malformation, hemophilia, spontaneous pneumothorax.

Case Report:

A 40 year old female, reported to emergency with complaints of dyspnea, palpitations and dry cough since last 7 days. She denied any associated history of hemoptysis, fever, chest trauma, and surgery. She had however similar breathlessness on and off since the last eight years. Patient had multiple hospitalizations for similar complaints and a significant past history with multiple first trimester abortions, repeated episodes of epistaxis and bleeding from finger tips. Patient was diagnosed and treated for RHD since last 2 years without any significant improvement. On General examination, pulse was 110 beats/minute, blood pressure 110/70 mmHg and respiratory rate of 22 breaths/minute. Her SpO, was 68% on room air. Grade III clubbing was noted in all the fingers with cyanosis(Figure 1). Nasal mucosa has very small multiple dilated vessels seen in right side of nasal septum in antero-inferior quadrant (Little's Area). Systemic respiratory examination was suggestive of massive pleural effusion on left side. Both the heart sounds were loud in intensity with tapping apex localized in 5 Inter-coastal space just lateral to mid clavicular line, pan systolic murmur present in mitral area radiating to axilla and back just lateral to spine. Routine ICU profile investigations were carried out to understand the cause of persistent hypoxia. Complete blood counts showed Haemoglobin of 7.9 gm%, platelet 80,000/mm^3 and total leucocyte count of 7,400/mm^3. Routine urine examination, renal and hepatic functions including coagulation profile were within normal limits. Arterial blood gas pH-7.46, PCO,-46 mmHg, PO,-37 mmHg and HCO,-32.3 mmol/L with oxygen saturation 67% Electrocardiogram showed sinus tachycardia. Chest X-ray showed opaque left hemithorax with shift of mediastinum to right (Figure 2). Right lung showed rounded opacities in lower zones. USG whole abdomen was suggestive of Cavernous transformation of portal vein with left side pleural effusion. Diagnostic pleurocentesis was carried out and pleural fluid was grossly haemorrhagic with haematocrit 34%, exudative in nature. Differential counts of pleural fluid showed 55% polymorphs and 45%Lymphocytes and cytology was negative for atypical or malignant cells. She remained hypoxic and cyanotic even after pleurocentesis and non invasive ventilation (Figure 3). A contrast-enhanced computed tomography of thorax was performed to know the state of underlying pleural space and lung parenchyma, CECT thorax was suggestive of multiple pulmonary arterio-venous malformation with feeding arteries from pulmonary artery and draining into pulmonary veins. Moderate left pleural effusion (hemothorax) with cardiomegaly and finding of Hepatic AVM(Figure 4).

Fig:1-Grade 3 Clubbing

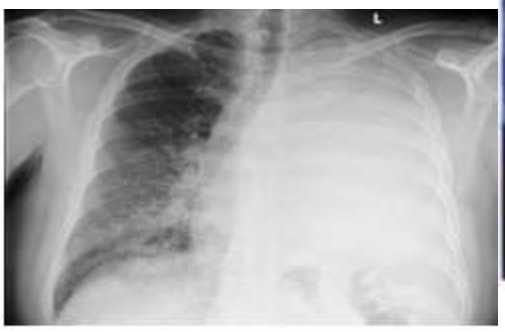

Figure:2 Chest Xray revealed massive effusion left with of mediastinum to opposite side

Figure:3 Post Pleurocentesis chest X ray revealed cannon balls shadowns in bilateral lower zone with left side costo phrenic angle blunting

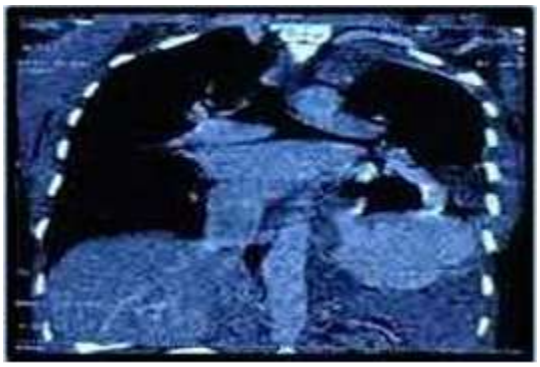

Fig4: CECT THORAX

MATERIALS AND METHODS :

This case study was conducted in the Department of Internal Medicine at Rama Medical College Hospital & Research Centre, Hapur, covering the study period from 22/06/2022 to 14/07/2023. Ethical approval was obtained from the institutional review board. Written informed consent was taken from the patient for publication of clinical details. The patient, a middle-aged adult, presented with complaints of progressive shortness of breath, easy fatigability, and intermittent episodes of hypoxemia. A detailed history, physical examination, laboratory evaluation, and radiological imaging were performed. History and Clinical Evaluation: A systematic approach was followed to elicit symptoms related to respiratory, cardiovascular, neurological, and hematologic systems. The patient reported worsening exertional dyspnea, which gradually progressed over several months. There was no history of recurrent epistaxis, cutaneous telangiectasia, or a family history suggestive of HHT. The absence of these features suggested the likelihood of a sporadic PAVM. No history of smoking, chronic lung disease, or cardiac illness was reported. On general examination, the patient was alert, oriented, and cooperative. Vital signs revealed mild resting hypoxemia with oxygen saturation fluctuating between 88–92% on room air. Mild cyanosis of the lips and nail beds was noted. No clubbing or significant lymphadenopathy was observed. Cardiovascular and abdominal examinations were unremarkable. Respiratory examination revealed diminished breath sounds in the right lower lung zone. Laboratory Investigations: Routine hematological and biochemical investigations were conducted, including complete blood count, liver function tests, renal function tests, and arterial blood gas (ABG) analysis. Hemoglobin levels were mildly elevated, likely due to compensatory erythrocytosis secondary to chronic hypoxemia. ABG analysis revealed low arterial oxygen tension. Coagulation profile was normal. Screening for autoimmune markers, viral serology, and inflammatory markers was conducted to rule out alternate

causes. All results were within normal limits. Radiological Investigations: A chest X-ray demonstrated a well-circumscribed opacity in the right lower lobe, suggestive of a vascular lesion. To confirm the diagnosis, a contrast-enhanced computed tomography (CECT) of the thorax was performed. Imaging revealed a single large arteriovenous malformation with a feeding artery arising from the right lower lobe pulmonary artery and a draining vein entering the pulmonary vein. The nidus of the malformation was well defined. No additional PAVMs were identified. No evidence of pulmonary embolism or interstitial lung disease was noted. Further Evaluation: A transthoracic echocardiogram (TTE) with contrast ("bubble study") was performed to assess the degree of right-to-left shunting. The study demonstrated early appearance of microbubbles in the left atrium, confirming the presence of a significant intrapulmonary shunt. Neurological examination was performed to rule out complications such as cerebral embolism. Magnetic resonance imaging (MRI) of the brain showed no abnormalities. Management: Based on imaging findings and the patient's symptomatic status, definitive management in the form of percutaneous transcatheter embolization was planned. The procedure was performed by an interventional radiologist under fluoroscopic guidance. A vascular access site was obtained through the right femoral vein. A catheter was advanced into the pulmonary arterial system, and selective angiography was performed to visualize the PAVM. The feeding artery was successfully occluded using detachable coils. Post-embolization angiography confirmed complete obliteration of the malformation with cessation of abnormal blood flow. Post-Procedure Care: The patient was monitored for potential complications, including bleeding, arrhythmias, and reperfusion injury. Oxygen saturation improved significantly following embolization, rising to 96% on room air. The patient was discharged after 48 hours with instructions for follow-up evaluation. Follow-Up: Regular outpatient follow-ups were conducted at 1 month, 3 months, and 6 months. Repeat chest imaging showed stable occlusion of the malformation with no residual or recurrent shunt. The patient's symptoms improved substantially, with resolution of dyspnea and normalization of exercise tolerance. No neurological complications were observed during follow-up. Data Compilation and Documentation: All clinical, laboratory, and imaging findings were systematically recorded. Data were analyzed qualitatively due to the descriptive nature of the case study.

Discussion:

Most commonly PAVMs are congenital (80%), out of these 47%-80% are associated with Osler-Weber-Rendu disease or hereditary haemorrhagic telangiectasia (HHT). On the other side, only 5%-15%of the HHT patient have a PAVM. Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu disease) is an autosomal dominant, systemic fibrovascular dysplasia in which telangiectasias, arteriovenous malformations, and aneurysms may be widely distributed throughout the body vasculature. Major clinical manifestations include: recurrent bleeding from mucosal telangiectasias and arteriovenous

malformations; hypoxemia, cerebral embolism, and brain abscess due to pulmonary arteriovenous fistulas; high-output congestive heart failure and portosystemic encephalopathy from hepatic arteriovenous malformations; and a variety of neurologic symptoms due to central nervous system angiodysplasia. Most deaths caused by PAVM are due to complications such as stroke, brain abscess, hemoptysis and hemothorax. Although the life expectancy is low in PAVM, the morbidity and mortality is mostly due the complications. Early diagnosis and management can reduce clinical manifestations and complications. Pulmonary haemorrhage due to spontaneous rupture of the PAVM is a potentially life-threatening complication that should be treated aggressively. There are two recommended therapeutic options in treatment of patients with PAVM: transcatheter embolisation and surgical resection (ligature, excision, segmentectomy, lobectomy, pneumonectomy), Lung transplantation is considered the last resort in the treatment and is indicated in failure of embolotherapy or failure of surgery, especially in patients with bilateral malformations. Considering that the patient had multiple PAVMs and severe hypoxemia, we adopted a conservative approach in treating this patient.

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