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Beyond Breathless: Unravelling The Enigma Of Pulmonary Arteriovenous Malformations

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Beyond Breathless: Unravelling The Enigma Of Pulmonary Arteriovenous Malformations

Abstract

Pulmonary Arteriovenous Malformations (AVMs) are abnormal connections between the pulmonary arteries and veins, leading to a direct shunting of blood without passing through the normal capillary bed. These AVMs can be associated with a rare genetic disorder called Hereditary Hemorrhagic Telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome. HHT is an autosomal dominant disorder characterized by the development of fragile telangiectasias in various organs, including the skin and mucous membranes. These telangiectasias are prone to bleeding, leading to recurrent nosebleeds and mucocutaneous bleeding. In patients with HHT, the most common site of AVMs is in the lungs. Pulmonary AVMs can cause significant health risks due to the right-to-left shunting of blood, leading to hypoxemia and possible complications like stroke, cerebral abscesses, and heart failure.

Keywords

Pulmonary Arteriovenous Malformations, Hereditary Hemorrhagic Telangiectasia, CT Pulmonary Angiography, Clubbing

BEYOND BREATHLESS : UNRAVELLING THE ENIGMA OF PULMONARY ARTERIOVENOUS MALFORMATIONS

A 15-year-old male patient presented with complaints of fever with chills and rigors, malena, coffee-coloured vomitus and epistaxis. Upon further inquiry, the patient disclosed a history of exertional dyspnea (MMRC Grade 1) since childhood. On physical examination, marfanoid habitus, palatal petechiae, central cyanosis and clubbing were noted.

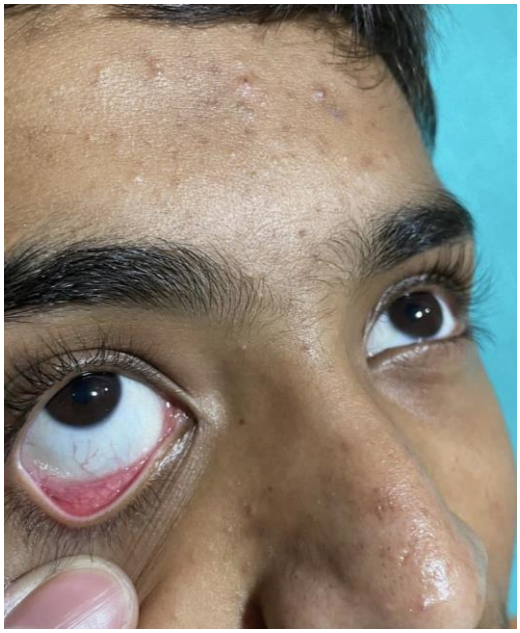


Figure 1.Conjunctival Telangiectasia

Figure 2.Clubbing

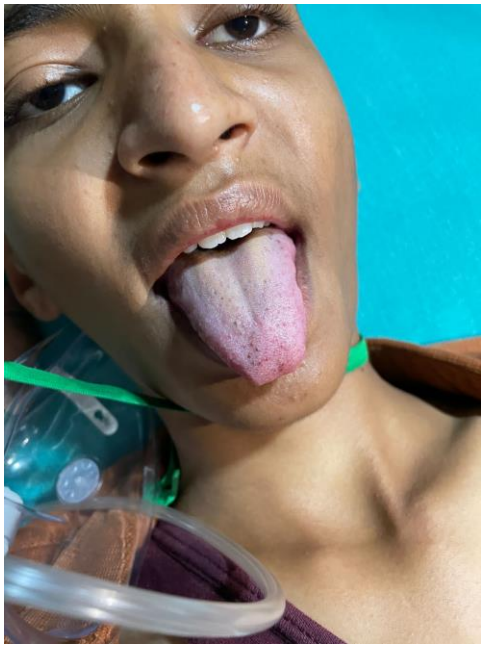


Figure 3.Bluish discolouration of tongue

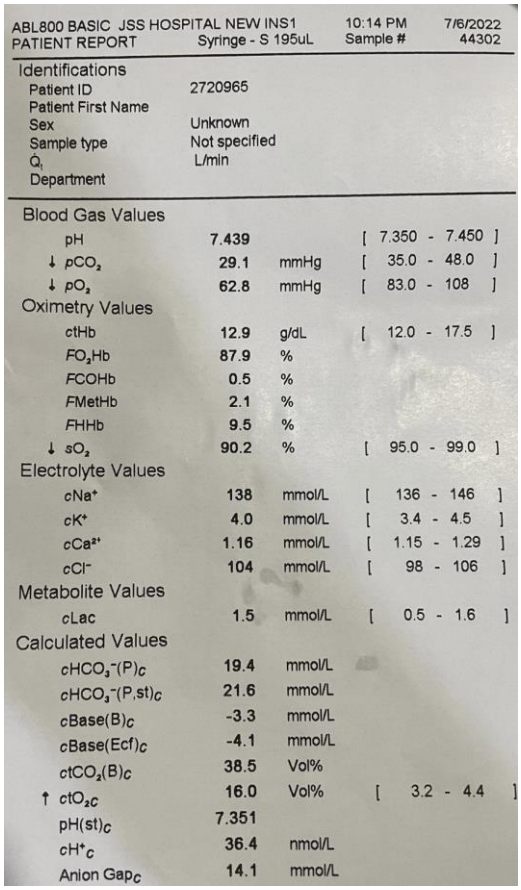
On cardiovascular system examination, visible pulsations were present in the 3rd and 4th intercostal space and apex beat was localized to 4th intercostal space. Auscultation revealed decreased air entry in bilateral axillary and scapular areas. Additionally a bruit

detected in the left subscapular region indicating a positive Suman's sign. On abdomen palpation, tenderness noted in the right hypochondriac and lumbar region, and a palpable liver.

INVESTIGATION:

The blood investigations showed polycythemia, blood smear showed microcytic hypochromic cells, reactive lymphocytes and thrombocytopenia.
Serology was positive for Dengue NS1 antigen and abdomen ultrasound revealed polyserositis and hepatomegaly.

The Arterial blood gas investigation showed low pCO2 and pO2 and minimally decreased oxygen saturation.



ABL800 BASIC JSS HOSPITAL NEW INS1 10:14 PM 7/6/2022
PATIENT REPORT Syringe - S 195uL Sample # 44302

Identifications			
Patient ID	2720965		
Patient First Name			
Sex	Unknown		
Sample type	Not specified		
Q_x	L/min		
Department			
Blood Gas Values			
pH	7.439		[7.350 - 7.450]
↓ pCO_2	29.1	mmHg	[35.0 - 48.0]
↓ pO_2	62.8	mmHg	[83.0 - 108]
Oximetry Values			
ctHb	12.9	g/dL	[12.0 - 17.5]
FO_2Hb	87.9	%	
FCOHb	0.5	%	
FMetHb	2.1	%	
FHHb	9.5	%	
↓ sO_2	90.2	%	[95.0 - 99.0]
Electrolyte Values			
cNa^+	138	mmol/L	[136 - 146]
cK^+	4.0	mmol/L	[3.4 - 4.5]
cCa^{2+}	1.16	mmol/L	[1.15 - 1.29]
cCl^-	104	mmol/L	[98 - 106]
Metabolite Values			
cLac	1.5	mmol/L	[0.5 - 1.6]
Calculated Values			
$cHCO_3^-(P)_c$	19.4	mmol/L	
$cHCO_3^-(P,st)_c$	21.6	mmol/L	
$cBase(B)_c$	-3.3	mmol/L	
$cBase(Ecf)_c$	-4.1	mmol/L	
$ctCO_2(B)_c$	38.5	Vol%	
↑ ctO_{2c}	16.0	Vol%	[3.2 - 4.4]
$pH(st)_c$	7.351		
cH^+_c	36.4	nmol/L	
Anion Gap_c	14.1	mmol/L	

Figure 4. Arterial Blood Gas Report

Chest X-ray had no markedly visible abnormalities and 2D-ECHO was normal. Hence was referred for further investigations.

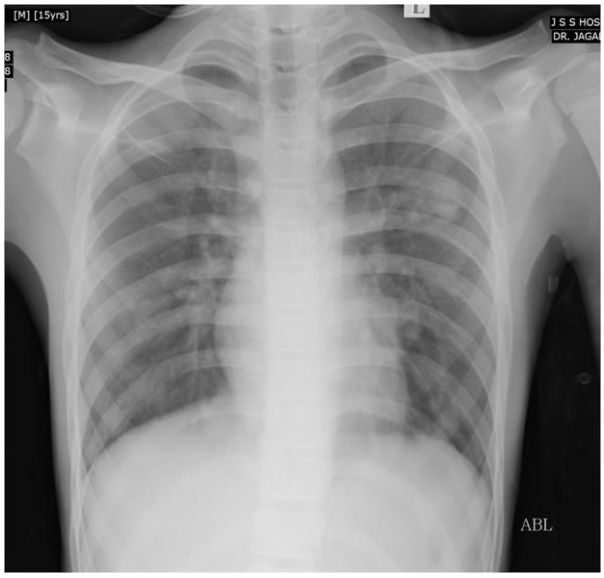
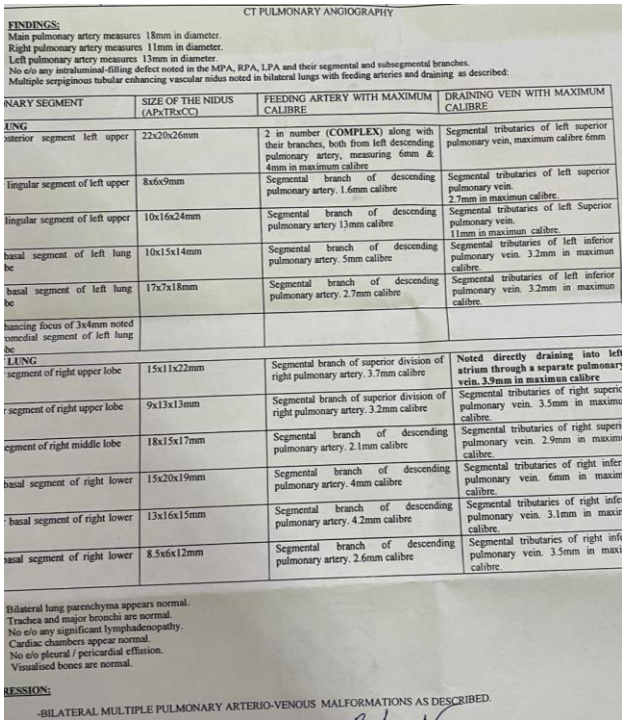


Figure 5. Chest X- ray



CT PULMONARY ANGIOGRAPHY

FINDINGS:

Main pulmonary artery measures 18mm in diameter.
Right pulmonary artery measures 11mm in diameter.
Left pulmonary artery measures 13mm in diameter.
No e/o any intraluminal-filling defect noted in the MPA, RPA, LPA and their segmental and subsegmental branches.
Multiple serpiginous tubular enhancing vascular nidus noted in bilateral lungs with feeding arteries and draining as described:

NARY SEGMENT	SIZE OF THE NIDUS (APxTRxCC)	FEEDING ARTERY WITH MAXIMUM CALIBRE	DRAINING VEIN WITH MAXIMUM CALIBRE
UNG			
osterior segment left upper	22x20x26mm	2 in number (**COMPLEX**) along with their branches, both from left descending pulmonary artery, measuring 6mm & 4mm in maximum calibre	Segmental tributaries of left superior pulmonary vein, maximum calibre 6mm
lingular segment of left upper	8x6x9mm	Segmental branch of descending pulmonary artery. 1.6mm calibre	Segmental tributaries of left superior pulmonary vein. 2.7mm in maximun calibre.
lingular segment of left upper	10x16x24mm	Segmental branch of descending pulmonary artery 13mm calibre	Segmental tributaries of left Superior pulmonary vein. 11mm in maximun calibre.
basal segment of left lung be	10x15x14mm	Segmental branch of descending pulmonary artery. 5mm calibre	Segmental tributaries of left inferior pulmonary vein. 3.2mm in maximun calibre.
basal segment of left lung be	17x7x18mm	Segmental branch of descending pulmonary artery. 2.7mm calibre	Segmental tributaries of left inferior pulmonary vein. 3.2mm in maximun calibre.
hancing focus of 3x4mm noted comedial segment of left lung be			
LUNG			
segment of right upper lobe	15x11x22mm	Segmental branch of superior division of right pulmonary artery. 3.7mm calibre	**Noted directly draining into left atrium through a separate pulmonary vein. 3.9mm in maximun calibre**
segment of right upper lobe	9x13x13mm	Segmental branch of superior division of right pulmonary artery. 3.2mm calibre	Segmental tributaries of right superio pulmonary vein. 3.5mm in maximu calibre.
egment of right middle lobe	18x15x17mm	Segmental branch of descending pulmonary artery. 2.1mm calibre	Segmental tributaries of right superi pulmonary vein. 2.9mm in maximu calibre.
basal segment of right lower	15x20x19mm	Segmental branch of descending pulmonary artery. 4mm calibre	Segmental tributaries of right infer pulmonary vein. 6mm in maxim calibre.
basal segment of right lower	13x16x15mm	Segmental branch of descending pulmonary artery. 4.2mm calibre	Segmental tributaries of right infe pulmonary vein. 3.1mm in maxi calibre.
basal segment of right lower	8.5x6x12mm	Segmental branch of descending pulmonary artery. 2.6mm calibre	Segmental tributaries of right infe pulmonary vein. 3.5mm in maxi calibre.

Bilateral lung parenchyma appears normal.
Trachea and major bronchi are normal.
No e/o any significant lymphadenopathy.
Cardiac chambers appear normal.
No e/o pleural / pericardial effusion.
Visualised bones are normal.

RESSION:

-BILATERAL MULTIPLE PULMONARY ARTERIO-VENOUS MALFORMATIONS AS DESCRIBED.

Figure 6. CT Pulmonary Angiography

In CT pulmonary angiography, multiple serpiginous tubular enhancing vascular nidus were noted in bilateral lungs with feeding arteries and draining vessels. This confirmed the presence of bilateral multiple pulmonary arteriovenous malformations(PAVM).

FINAL DIAGNOSIS:

Based on the relevant reports the diagnosis was made as dengue with suspected Hereditary Hemorrhagic Telangiectasia(HHT).

The patient was managed symptomatically for dengue and was referred to interventional cardiology to undergo a percutaneous trancatheter embolization with a follow up request after 6 months.

DISCUSSION:

Pulmonary Arteriovenous Malformation (AVM) is a rare clinical problem with an incidence of 1 in 50,000 individuals. It involves an abnormal communication between a pulmonary artery and a pulmonary vein, bypassing the pulmonary capillary bed. While most cases are congenital, some may also be acquired. Notably, most patients with pulmonary AVMs have hereditary hemorrhagic telangiectasia (HHT), an autosomal dominant disorder characterized by cutaneous telangiectasia, a family history of the disorder, and recurrent epistaxis.
These AVMs are typically seen as unilateral lesions in the subpleural space of the lower lobes. The primary defect results in right-to-left shunting, leading to a mixture of deoxygenated and oxygenated blood, causing hypoxemia that persists despite the administration of 100% oxygen. Patients commonly present with epistaxis, dyspnea on exertion, platypnea-orthodeoxia, clubbing, cyanosis, mucocutaneous telangiectasias, and audible murmurs or bruits over the location of large pulmonary AVMs.

Radiologically, pulmonary AVMs appear as round masses of uniform density on chest radiographs, usually lobulated and sharply-defined. Additional investigations, such as Contrast-enhanced computed tomography (CT) and pulmonary angiography, are crucial for confirming the diagnosis.

Percutaneous embolization is the primary therapy for pulmonary AVMs, with a success rate of 98%. The prognosis after treatment is generally favorable. However, untreated pulmonary AVMs can lead to significant morbidity and mortality, including cerebral abscess, stroke, and potentially life-threatening pulmonary AVM arterial rupture.

The differential diagnosis for patients with pulmonary AVMs is broad, making the patient's initial presentation essential in guiding the evaluation. Regular follow-up is imperative following treatment to monitor for any recurrence or complications.

CONCLUSION:

Pulmonary arteriovenous malformation is a rare but significant clinical problem associated with hereditary hemorrhagic telangiectasia. This case is of paramount importance to the medical fraternity because of the need for an interprofessional approach in diagnosis of HHT with PAVM. It is often underdiagnosed due to a lot of asymptomatic cases and broader differentials. Early identification and diagnosis can prevent rapid disease progression and higher complication rates, giving a broader horizon of management options to prevent a life-threatening catastrophe.

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