Content available at: https://www.ipinnovative.com/open-access-journals

IP Indian Journal of Immunology and Respiratory Medicine

Journal homepage: https://www.ijirm.org/

Case Report

An unusual case of hemoptysis secondary to Abernethy malformation type II and hepatopulmonary syndrome, pulmonary arterial hypertension

Shaik Umar Pasha[1], Shaik Abdul Waseem[1], Nagarjuna Reddy Sunkara[1], Seenu Ayyakannu[1], Ramakrishna Rachakonda[1,*]

[1] *Dept. of Pulmonary Medicine, NRI Medical College, Chinnakakani, Guntur, Andhra Pradesh, India*

ARTICLE INFO

Article history:
Received 29-10-2021
Accepted 22-12-2021
Available online 31-12-2021

Keywords:
Abernethy Malformation
Hepatopulmonary syndrome
Extrahepatic shunt
Pulmonary arterial hypertension

ABSTRACT

A twenty years old male college student came with history of hemoptysis. His chest X-ray showed diffuse infiltrative shadows and he was diagnosed as a case of a case of pulmonary tuberculosis with hemoptysis. On detailed investigations he was found to have Abernethy malformation Type-II with primitive portal vein joining extrahepatic inferior vanacava leading to cirrhosis of liver, porto-pulmonary syndrome, pulmonary arterial hypertension and hemoptysis.

1. Introduction

An abnormal communication between portal vein and hepatic vein results in a portosystemic shunt which results in hepatopulmonary syndrome with resultant porto pulmonary communication and pulmonary arterial hypertension. Here we are presenting this abnormal communication in a twenty years old boy. Normally pulmonary tuberculosis or vasculitis are common causes of hemoptysis. This is a unique case where this congenital abnormality is detected in a twenty years old male patient. This is a rare congenital abnormality diagnosed in a slightly older age group compared to the other case reports which were reported in younger children.

A twenty years old male patient, a college student was brought to the department of Respiratory Medicine NRI Medical College in March 2019 with symptoms of cough with expectoration and hemoptysis.

Patient had multiple bouts of hemoptysis amounting to 30ml to 50ml. He was seen by a local chest physician. CT scan chest showed multiple areas of ground glass opacities and patchy consolidation in both middle and lower lobes in both lungs. Patient was given anti tuberculosis drugs for about 10 days.

In our tertiary care centre this patient was initially thought as a case of vasculitis and hemoptysis and was given corticosteroid therapy

2. Case Report

Patient was tall 198cm in height with a Span of 178cm. He had no high arched palate. He had clubbing of fingers and toes. Gynecomastia was present. Body was hair sparse. There was right testicular atrophy. There was genu valgum deformity of knees.

2.1. Summary of investigations done

HRCT Chest done in December 2019 suggested cardiomegaly with dilated pulmonary trunk and bilateral prominent pulmonary arteries, bilateral plethoric lung segments–Possible pulmonary arterial hypertension. There was evidence of interstitial lung disease and airspace consolidation.

* *Corresponding author*.
E-mail address: ramakrishna45@yahoo.co.in (R. Rachakonda).

https://doi.org/10.18231/j.ijirm.2021.051

CECT chest suggested alveolar hemorrhage with confluent consolidations with surrounding ground glass opacities in bilateral upper lobes (Likely infective consolidations). Mild cardiomegaly with mild dilated right ventricles. Main pulmonary artery was dilated with a diameter of 35 mm. Diameter of left pulmonary artery was 24 mm and that of right pulmonary artery was 24 mm.

Ultrasound examination of abdomen suggested coarse echo texture of liver with prominent inferior vanaca, hepatic veins and portal vein and possible cardiac cirrhosis.

Hepatic venous Doppler showed chronic liver disease with multiple peripancreatic collaterals. Portal vein at hilum was normal, but the intrahepatic portal vein was attenuated. Right and left hepatic veins were not visualized. Main hepatic vein was normal. There was no evidence of intrahepatic collaterals

Upper GI endoscopy was normal. There was hiatus hernia, Hill's Grade II. There were no esophageal or fundal varices.

2.2. Radio imaging showed hypertrophied and tortuous hepatic artery

Bronchoscopy revealed bronchomalacia of right main bronchus with about 20-30% narrowing of right main bronchus lumen. Rest of the bronchial tree was normal. No endobronchial lesion was seen. Serial aliquots did not show increase in haemorrhagic returns. BAL was taken from both the upper lobes. Transbronchial lung biopsy was deferred due to nasal bleed and poor patient cooperation

2.3. CTPA

Revealed main pulmonary artery was dilated and measured around 38mm in diameter. Right and left pulmonary arteries and their branches also appear dilated. There was no evidence of pulmonary embolism upto segmental artery level. There was right ventyricular hypertrophy, RA and RV dilatation. There was no evidence of pericardial effusion. Hypertrophy of bronchial arteries was seen.

2.4. Lungs and pleura

There are focal areas of consolidation with surrounding ground glass densities in the left upper lobe and anterior segment of right upper lobe. Mild interlobular septal thickening was also seen in both lungs. Few tortuous vessels with suspicious tiny AV fistulas were seen in the right lower lobe and even smaller ones elsewhere in both lungs. Trachea and main bronchi were normal.

There were sub centimeter hilar nodes on both sides. There was a healed fracture of right clavicle. CT Abdomen and pelvis showed findings of portosystemic shunt between the main portal vein and intrahepatic IVC consistent with Abernathy malformation type II. The right portal vein branch was attenuated. The left branch was not visualized .Few prominent vessels were seen in bilateral lung bases. There were no focal lesions in liver. There was a porto systemic shunt with communication of main portal vein and intrahepatic IVC near porta. The main portal vein measured 17mm. spleen was normal. There were no radiopaque calculi in the gall bladder. Pancreas and adrenals were normal.

Echo showed normal LV systolic function with no regional wall motion abnormality. There was mild tricuspid regurgitation, mild PAH and mildly dilated RA. Pulmonary valve was displaced anterior to Aortic valve in the RVOT side in aortic SAX View. Right Ventricular systolic function was normal.

2.5. Other Investigations done were- Anti-Nuclear antibody

Negative. Prothrombin time 14.1 Sec. INR: 1.18. LDH 604 U/L (Normal 230 to460 U/L: Hb 15.9 g/dl.RBC 5.6 million/cubic mm. WBC 10900/cubic mm. Platelet count 1.6 Lakhs. PCV 54%. MCV 95 Cubic microns. MCH 26 picograms. MCHC 28%. RDW 13.5%. Differential Counts- Neutrophils 82%, Lymphocytes 14% Eosinophils 01%, Monocytes 03%. Peripheral smear was normal. ESR 10mm.hr. Serum bilirubin Total 3.0 mg%, Direct 0.5, Indirect 2.5 mg%, urea 16, serum creatinine 0.8 mg/dl. Serum cortisol 3.48 micrograms/dl. Low AST 19 U/L, ALT 40 U/L Alkaline Phosphatase 67 U/L. Serum Proteins 5.5g/L albumin 2.7g/L, globulin 2.8g/L, urea 17 mg%, sodium 138 mmol/L, potaassium 4.1 mmol/L. GeneXpert of sputum for tuberculosis was not suggestive of MTB. BAL Negative for fungal culture. BAL fluid Negative for Mycobacterial Culture. BAL smears revealed haemosiderophages.

Twenty years old male patient has presented with hemoptysis and was found to have chronic liver disease dilated portal vein, chronic hemolytic anemia, hemosiderosis, hepato pulmonary syndrome, findings of portosystemic shunt between the main portal vein and intrahepatic IVC consistent with Abernathy malformation type II, pulmonary arterial hypertension, pulmonary arterio-venous communications with recurrent intrapulmonary hemorrhage with fleeting shadows in CT chest. Before publishing the data written informed consent from the patient and the family members was taken.

3. Discussion

Abernethy malformations are a rare vascular anomalies. They consist of persistent embryonic vessels causing congenital Porto systemic shunts.

In type I Abernethy malformation splenic vein and superior mesenteric vein open separately in to IVC. This abnormality is usually seen in females.

In Type II Abernethy malformation superior mesenteric vein and splenic vein form a common trunk before draining into IVC.

Our case is a unique case because it is presented in a male patient in the third decade of life. There were features of cirrhosis of liver with left testicular atrophy, sparse hair on the body. These features accompanied a congenital vascular anomaly of liver vessels with primitive portal vein opening in to intra-hepatic inferior vanacava. This resulted in porto-pulmonary hypertension and tortuous pulmonary vessels abnd resultant alveolar hemorrhage and hemoptysis. There was indirect hyperbilirubinemia because of hemolysis.

These vascular anomalies were reported world over but a majority of them were reported among female children.

There are several case reports of similar abnormality. Pathak et al., presented this abnormality in a five year old female child who presented with a short history of jaundice and Abernethy 1b malformation. This case was diagnosed with ultrasound, Doppler and CT-angio studies. They suggested early closure of the shunt before complications develop.[1] Abernethy malformation could lead to hepatopulmonary syndrome because of bypassing the liver metabolism and release of toxic substances in to the systemic and pulmonary circulatuions.[2] Congenital extrahepatic porto systemic shunts are best detected by MDCT (Multi detector Computerized tomography).[3] Extrahepatic porto systemic shunts are responsible for hepato pulmonary syndrome and hypoxic decompensation can be prevented by early closure of the shunt.[4] In our case hypoxic decompensation was not present despite higher age of presentation to the hospital.

In a case report by Sukru et al., a case of type Ib abnormality was presented in a nine year old girl whose orthodeoxia responded to auxiliary partial orthotopic liver transplantation (APOLT) two months after surgery.[5]

A similar case of Type II Abernethy malformation was reported in pediatric age group by Kimberly Saulters with hepatopulmonary syndrome. This HPS can lead to hyperammonemia and hepatic encephalopathy.[6]

As seen in an Indian study Abernethy malformation with porto systemic shunt can be completely asymptomatic to a highly symptomatic states like hepato-pulmonary syndrome, severe pulmionary arterial hypertension and can even cause hepatic carcinoma.[7]

A similar case like ours was reported in a 19 year old male by Xue-Yan et al., with cyanosis and dyspnea caused by extrahepatic porto systemic shunt and pulmonary arterial hypertension.[8]

Abernethy malformation with hepatopulmonary syndrome can rarely lead to hepato cellurar carcinoma as presented by Benedict et al.[9]

Early orthodeoxia was presented in a seven year old boy with Abernethy malformation and hepatopulmonary syndrome by Maria Joana Osario et.al.[10]

Abernethy malformation is a rare congenital vascular anomaly that causes extra hepaticporto systemic shunt that can lead to pulmonary arterio venous malformations, pulmonary arterial hypertension, orthodeoxia, respiratory failure and even hepato cellular carcinoma. In our patient it came to the diagnosis through presentation of massive hemoptysis and alveolar haemorrhage. Early closure of the shunt and orthotopic liver transplantation can help in improvement of symptoms.

4. Acknowledgements

We acknowledge the services of the department of Radiology and Imaging of NRI Medical College, Department of Pulmonology NRI Medical College, Department of Gastroenterology, NRI Medical College, Guntur.

5. Conflict of Interest

The authors declare no conflict of interest.

6. Source of Funding

None.

References

1. Pathak A, Agarwal N, Mandliya J. Abernethy malformation: a case report. *BMC Pediatr*. 2012;12:57. doi:10.1186/1471-2431-12-57.
2. Alvarez AE, Ribeiro AF, Hessel G, Baracat J, Ribeiro J. Abernethy malformation: one of the etiologies of hepatopulmonary syndrome. *Pediatr Pulmono*. 2002;34(5):391–4.
3. Ghuman SS, Gupta S, Buxi T, Rawat KS, Yadav A, Mehta N, et al. The Abernethy malformation-myriad imaging manifestations of a single entity. *Indian J Radiol Imaging*. 2016;26(3):364–72.
4. Sahu MK, Bisoi AK, Chander NC, Agarwala S, Chauhan S. Abernethy syndrome, a rare cause of hypoxemia: A case report. *Ann Paedatr Cardiol*. 2015;8(1):64–6.
5. Emre S, Arnon R, Cohen E, Morotti RA, Vaysman D, Shneider BL, et al. Resolution of hepatopulmonary syndrome after auxiliary partial orthotopic liver transplantation in Abernethy malformation. A case report. *Liver Transpl*. 2007;13(12):1662–8.
6. Saulters K. Hepatopulmonary Syndrome in the Presence of Abernethy Malformation: A Pediatric Case Report. *J Paediatr Health Care*. 2015;29(1):104–7.
7. Azad S, Arya A, Sitaraman R, Garg A. Abernethy Malformation , our experience from a tertiary Cardiac care Center, Rdeview of Literature. *Ann Paed Cardiol*. 2019;12(3):240–7.
8. Xue-Yan D, Feng C, Xian XZ, Hong W, Shao-Ping C, Yongowen Q, et al. A rare cause of Cyanosis: Hepato Pulmonary syndrome caused by Caused by Congenital Extra hepatic porto systemic shunt. *Case Rep Vasc Med*. 2011;p. 508171. doi:10.1155/2011/508171.
9. Benedict M, Rodriguez-Davalos M, Emre S, Walther Z, Morotti R. Congenital Extrahepatic Portosystemic Shunt (Abernethy Malformation Type Ib) With Associated Hepatocellular Carcinoma: Case Report and Literature Review. *Pediatr Dev Pathol*. 2017;20(4):354–62. doi:10.1177/1093526616686458.
10. Osorio MJ, Bonow A, Bond GJ, Rivera MR, Vaughan KG, Shah A. Abernethy malformation complicated by hepatopulmonary syndrome and a liver mass successfully treated by liver transplantation. *Pediatr Transplant*. 2011;15(7):E149–51.

Author biography

Shaik Umar Pasha, Final Year Post Graduate Student

Shaik Abdul Waseem, Final Year Post Graduate Student

Nagarjuna Reddy Sunkara, Post Graduate

Seenu Ayyakannu, Post Graduate

Ramakrishna Rachakonda, Professor and HOD

Cite this article: Pasha SU, Waseem SA, Sunkara NR, Ayyakannu S, Rachakonda R. An unusual case of hemoptysis secondary to Abernethy malformation type II and hepatopulmonary syndrome, pulmonary arterial hypertension. *IP Indian J Immunol Respir Med* 2021;6(4):237-240.