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IDIOPATHIC PULMONARY ARTERIOVENOUS MALFORMATION - UNCOMMON PRESENTATION OF AN UNCOMMON DISEASE

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Abstract

Idiopathic pulmonary arteriovenous malformation - uncommon presentation of an uncommon diseases

Introduction

Pulmonary arteriovenous malformations (PAVMs) are rare pulmonary vascular anomalies that can be associated with a wide spectrum of clinical manifestations, some of which include life-threatening hemorrhage. We report a case of a patient with idiopathic PAVM presenting with an unusual complication - spontaneous hemothorax.

Case Report

A non-smoker 45-year old woman presented with left-sided chest pain and breathlessness over 7-day duration, with no history of chest trauma. She had no signs of hemodynamic instability and did not appear to be in significant respiratory distress with only mild partial respiratory insufficiency. Laboratory findings were within

normal limits apart from mild normocytic anemia and elevated D dimers. Chest X-ray confirmed left-sided pleural effusion. An emergency MSCT angiography showed no signs of pulmonary embolism but instead a probable AV malformation 17 mm in diameter was shown in left lingular area. Diagnostic thoracocentesis revealed hemorrhagic exudate in organizing stage. After thoracic drainage was performed, along with antibiotic treatment, gradually regression of hemothorax occurred. MSCT angiography performed three months later then confirmed a simple PAVM in the peripheral lingular area supplied from superior left pulmonary artery, and with a drainage vein entering the pulmonary veins and left atrium. The feeding artery with the diameter of 4.5-6mm was found to be suitable for transcatheter embolization, which was afterwards successfully performed with six embolization coils.

Conclusion

Most PAVMs are associated with HHT and only a small portion of PAVMs is believed to be idiopathic. Hemothorax as a presenting feature of PAVM is a very rare occurrence. There is little data on idiopathic PAVMs in the literature, especially presenting with hemothorax, and hopefully this case could benefit treatment some of PAVM patients. We have shown that management of PAVM related hemothorax initially by thoracic drainage followed by later on performed catheter embolisation of the PAVM in a low risk patient led to a successful outcome.