Pulmonary Arteriovenous Malformations Presenting With Hypoxemia

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INTRODUCTION

An 81 year old female with a past medical history of upper GI bleeds, recurrent epistaxis, transient ischemic attacks, anemia and bradydysrhythmia requiring pacemaker was admitted to the hospital for an acute lateral wall myocardial infarction. Five days later, the patient was noted to have persistent hypoxemia to 83% on room air. She complained of shortness of breath while sitting upright and favored the supine position. She had no other significant pulmonary complaints.

The patient has a family history of recurrent epistaxis. Social history is negative for tobacco use, alcohol abuse or exposure to toxins.

DIAGNOSTIC STUDIES

ABG: initial ABG revealed low O2 saturation

VQ lung scan: ruled out pulmonary embolism; scintigraphy showed uptake of technetium over the brain and kidneys suggesting right to left shunting of blood

Spirometry: ruled out underlying restrictive or obstructive pulmonary pathology

ECHO with doppler: revealed preserved left ventricular ejection fraction with moderate mitral regurgitation, no septal defects or intracardiac shunting

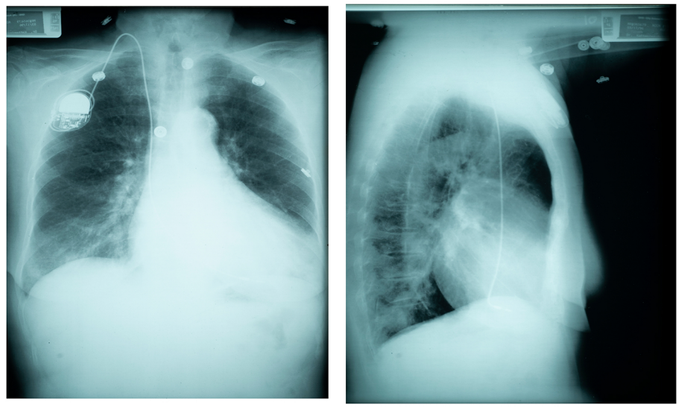
Chest X-ray PA/Lateral: showed cardiomegaly, generalized vascular prominence, and a permanent pacemaker. Lateral view suggested a tortuous, tubular density at the left lung base

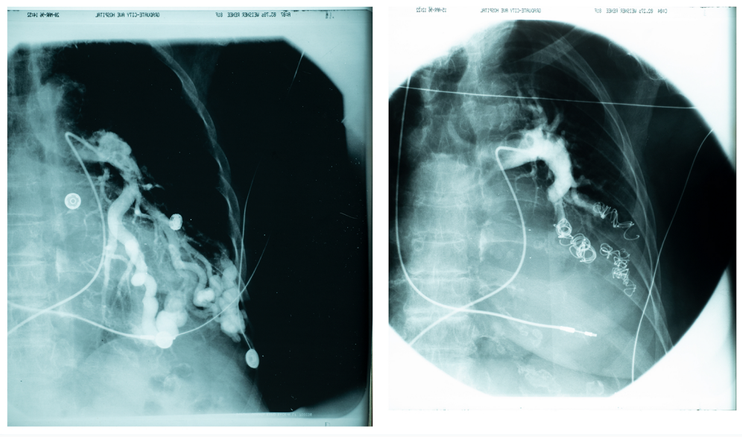
DIAGNOSTIC STUDIES

High resolution CT scan: improved visualization of abnormalities; revealed nodular tortuous densities of the left lung base consistent with pulmonary arteriovenous malformations (AVMs)

Pulmonary angiography: test of choice, confirmed presence of AVMs and demonstrated resolution of AVMs after successful percutaneous transcatheter embolotherapy

IMAGING





RESULTS

The patient was found to have multiple AVMs at the left lung base which were successfully treated with percutaneous transcatheter embolotherapy. These pulmonary abnormalities in conjunction with her past medical history, lead to the diagnosis of Osler-Weber-Rendu disease (OWRD). This rare autosomal dominant condition causes vascular dysplasia throughout the body and has a high propensity for bleeding. The patient was discharged from the hospital with improved oxygen saturation of 92% on room air after coil embolization of her AVMs.

CONCLUSION

This 81 year old hypoxic female with pulmonary AVMs associated with recurrent epistaxis is consistent with hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu disease. Her pulmonary abnormalities were successfully treated with percutaneous coil embolization.

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