BRIEF OVERVIEW OF PULMONARY ARTERIOVENOUS MALFORMATIONS

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Pulmonary arteriovenous malformations (PAVM) generally involve an abnormal vascular conduit between a pulmonary artery and vein; thereby, producing an intra-pulmonary right-to-left shunt. Mayo Clinic series over the last 4 to 5 decades indicate an incidence of 4 cases annually, more commonly in women than men. Clinical presentations include dyspnea, platypnea, and hemoptysis. As most PAVM are in association with hereditary hemorrhagic telangiectasia (HHT), the patient may also present with epistaxis or gastrointestinal bleeding. Clinical signs include telangiectasias, digital clubbing, cyanosis, orthodeoxia, and anemia. Complications, in addition to those mentioned above, include cerebral embolism and abscess. Chest x-ray often shows a lesion consistent with a PAVM or be interpreted as simply a pulmonary nodule. Chest CT is more sensitive and if performed with contrast, specific as well. While saline contrast echocardiography is often used to screen for right-to-left shunting, quantification requires either a shunt study with 100% oxygen or radionuclide imaging. Advances with percutaneous intravascular embolization have replaced surgery for PAVM ≥ 3 mm. Hepatopulmonary syndrome (HPS) should also be mentioned and may be due to discreet PAVM. More often, HPS is due to basilar vasodilation and relative right-to-left shunting that may improve with high flow oxygen and is definitively treated with orthotopic liver transplantation. Antibiotic prophylaxis is recommended for procedures associated with bacteremia. Interestingly and counterintuitively, both HHT and HPS can present with pulmonary arterial hypertension.

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