

Background:

Pulmonary arteriovenous malformations (PAVMs) are abnormal communications between pulmonary arteries and veins. Most PAVMs are idiopathic but part of them are congenital and due to hereditary hemorrhagic telangiectasia (HHT, also called Osler-Weber-Rendu syndrome). The clinical features suggestive of PAVMs are stigmata of right-to-left shunting (dyspnea, hypoxemia, cyanosis, cerebral embolism, brain abscess), unexplained hemoptysis, or hemothorax. For most patients with suspected PAVMs, the initial test of choice to evaluate the presence of a right-to-left shunt is saline bubble contrast echocardiography (TTCE, also known as “bubble echocardiography”). The other diagnostic test is chest CT scan. The feeding artery diameter, PAVM-related symptoms, and the patient’s ability to tolerate the procedure are the most important factors for selection of patients candidate for treatment. When indicated, most patients are treated with embolotherapy. Surgery treatment is another option in case of embolization failure.



Image 1: Angio-TC scan of the chest showing MAV

Case report :

a 19 years old man presented to Emergency Department complaining dyspnea, polycythemia (Hb 19.8 gr/dl, Htc 59%) and persistent hypoxemia (SaO₂ <85%) in course of supplemental oxygen therapy. In her medical history only autoimmune thyroiditis. The clinical exam revealed clubbing fingers. The arterial blood gas test showed hypoxemic respiratory failure with respiratory alkalosis (pH 7.45, pO₂ 46 mmHg, pCO₂ 29 mmHg, SpO₂ 83%, HCO₃ 22.5 mmol/l). Metabolic panel, thoracic x-rays, electrocardiogram and echocardiography were all normal. Angio-CT scan of the chest detected multiple PAVMs in subsegmental arteries and veins of the left lung with marked dilatation of the left pulmonary vein. TTCE showed severe PAVM with right-to-left shunt. The patient was treated with injection of embolic material by selective angiographic catheterization of the biggest PAVMs (embolotherapy). The procedure was repeated after a few months to complete the closure of remaining PAVMs. The patient subsequently was in good general conditions, asymptomatic for dyspnea, with persistent mild desaturation on exertion. Other tests were made for detecting other PAVMs associated to HHT, in particular in splanchnic and cerebral districts, which were normal.



Image 2: selective angiographic catheterization

Conclusion & perspectives :

PAVMs are uncommon in the general population, but they need to be considered in differential diagnosis of common pulmonary sign and symptoms such as hypoxemia, pulmonary nodules and hemoptysis. PAVMs are often associated with malformations in other body districts which need to be evaluated in case of suspected HHT syndrome. PAVMs morbidity and mortality are related to development of serious complications including stroke, brain abscess, chronic hypoxemic respiratory failure and life-threatening hemoptysis or hemothorax.