Severe hypoxemia due to intrapulmonary right-to-left shunt: AVM embolization through internal jugular access

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Introduction
Respiratory hypoxia is a common cause of in-patient deterioration. Although the most frequent causes are ventilation-perfusion mismatch or hypoventilation, a careful history, examination and extended differential diagnosis are invaluable especially in the presence of hypoxia refractory to supplemental oxygen.

Pulmonary arteriovenous malformation (PAVM) is an abnormal communication between the pulmonary arteries and veins and is a rare cause of refractory hypoxia due to intrapulmonary right-to-left shunt is a rare cause of hypoxia. Although it may occur as a primary disease in the lung, usually it is associated with hereditary hemorrhagic telangiectasia (HHT), formerly known as Rendu-Osler-Weber syndrome. Main clinical features of this autosomal dominant disease are epistaxis, mucocutaneous telangiectasias, and AVM (lung, liver or brain). In addition, there is an increased incidence of venous thromboembolism (responsible for cerebrovascular events in these patients).

We present a case of unexplained hypoxia and neurological deficits caused by a PAVM and its approach. To our best knowledge this was the first successively percutaneous transcatheter embolization of a PAVM through internal jugular vein access in a patient with absent infrarenal inferior vena cava.

Case report
A 55-year-old woman was admitted in the Internal Medicine ward due to acute respiratory distress presented with dyspnea, hypoxemia, sudden paraparesis (lower limb, graded 3/5) and hypertensive crisis. Vital signs at admission: HR 103 bpm, RR 25 cpm, BP 197/98 mm Hg, oxygen saturation of 88% on room air. Blood gas analysis (FiO2 50%) showed pH 7.45, PaCO2 27 mm Hg, PaO2 61 mm Hg, HCO3− 21.9 meq/L and a PaO2/FiO2 ratio of 122. No other remarkable results showed on her laboratory work-up (including cultures of blood, urine, stool, bronchial secretions, and CSF).

Despite supportive treatment with noninvasive ventilation and bronchodilation, she continued to deteriorate and required intubation and invasive mechanical ventilation due to persistent hypoxemia and was admitted to the ICU. Pursuing an etiological diagnosis, a neuroaxis magnetic resonance imaging was requested to clarify the lower limb paresis which showed a left parieto-occipital acute ischemic lesion; an echocardiogram was done to exclude intracardiac right-to-left shunt; and finally, a contrast-enhanced computed tomography scan in venous phase of the chest, abdomen, and pelvis was done to search for extracardiac shunts. The scan revealed a 35-mm PAVM within left upper lobe in a patient with absent infrarenal inferior vena cava and common iliac veins (Fig. 1). The femoral veins were draining into external iliac veins which in turn were draining into the ovarian veins.

The patient was referred to the Interventional Radiology Unit and proposed to percutaneous transcatheter embolization of the PAVM (assumed as cause for both persistent hypoxemia and embolic stroke lesion).

For the intervention, an ultrasound-guided puncture of the right internal jugular vein was done and a 6-Fr introducer-sheath deployed. This alternative access (commonly made through the femoral vein) was used because the vascular anatomical features found in the contrast-enhanced computed tomography (Fig. 1). The feeding artery was selected with a cobra 1 5-Fr diagnostic catheter and 0.035” hydrophilic guidewire. Over a stiff hydrophilic guidewire, a 6-Fr guide catheter was advanced to the distal area of the feeder vessel. Percutaneous transcatheter embolization of AVM was performed with a 12-mm Amplatzer vascular plug II (Fig. 2).

Immediately after the embolization, the patient vitals started to improved with reduced heart rate (73 bpm) and oxygen saturation rising to 100% and the blood gas analysis showing correction of the hypoxemia with (FiO2 50%) pH 7.308, PaO2 141.1 mm Hg, PaCO2 44.7 mm Hg, HCO3− 21.9 meq/L and a PaO2/FiO2 ratio of 282. No immediate complications occurred during the procedure and the patient was readmitted in the ICU. The patient was extubated shortly after embolization and the respiratory symptom vanished. The neurologic symptoms, however, persisted and a multimodal physical therapy program addressing lower limb weakness, mild dysphagia for liquids, and dysarthria was implemented.

Discussion
PAVM is a benign condition yet presenting several times as refractory hypoxia due to intrapulmonary right-to-left shunt. Depending on the size and number of PAVM presented and the degree of shunt the clinical presentation can vary from asymptomatic (despite altered gas exchange as noted in blood gas analysis) to severe hypoxia with no correction with high
Figure 1. Axial section (A) and sagittal MIP reconstruction (B) revealed a left upper lobe arteriovenous malformation (AVM) with a single feeding artery (8-mm caliber) originating from the pulmonary artery and a single draining vein. Axial sections at the level of infra renal aorta (C) and coronal mean MIP reconstruction (D) shows nonvisualization of IVC below renal veins, enlarged ovarian veins (∗) and absent common iliac veins. MIP = maximum intensity projection.

Figure 2. Status pre- (A) and postembolization (B) of the left pulmonary artery segmental branch, with arteriovenous malformation occlusion.
inspired fraction of oxygen. Contrast-enhanced computed tomography is the criterion standard tool for diagnosis and mandatory to plan any intervention. Percutaneous transcatheter embolization is the preferred treatment because it avoids general anesthesia and the risk of a major surgery. To our best knowledge, this is the first that an embolization of AVM is completed through an internal jugular access.

Concerning the diagnosis of HHT, consensus criteria are based on the presence of spontaneous epistaxis, mucocutaneous telangiectasias, AVM, and a positive familial history. Definitive diagnosis is attained when 3 criteria are met, with 2 criteria making the diagnosis possible. Our patient has a probable diagnosis of HHT with 2 criteria present: PAVM and family history positive – her son was diagnosed based on 3 criteria (epistaxis, telangiectasias, and AVM) few weeks later.

References

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