A REVERSIBLE CAUSE OF CYANOSIS

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A 20-year-old male was referred to our institute for evaluation of easy fatigability and breathlessness. On physical examination, he had central cyanosis with 78% SPaO₂ and clubbing. There were bilateral symmetrical pulses and cardiac examination was normal. Chest x-ray revealed homogenous tubular opacity in the left upper zone. Electrocardiogram (ECG) and two-dimensional echocardiographic study were normal. Contrast echocardiography was done by injecting agitated saline in the upper limb peripheral vein. Immediate (within 3 cardiac cycles) appearance of microbubbles in the LA confirmed the provisional diagnosis of pulmonary arteriovenous fistula (PAVF). Computed tomography of pulmonary angiography revealed a large PAVF arising from left pulmonary artery (Figure 1). There were no other features suggestive of hereditary hemorrhagic telangiectasia. Since patient was symptomatic with desaturation, he was taken for cardiac catheterization with intent to closure of the fistula. Selective left pulmonary artery (LPA) angiography with 6 Fr Pigtail revealed a large PAVF, draining into the LA via a large vertical tubular vessel. A 6 Fr Judkins right (JR) catheter was advanced over the guidewire into the sac through the largest arterial feeder. The targeted arterial feeder measuring 15 mm was occluded by deploying a 18 x 20 mm duct occluder (Cocoon duct occluder, Vascular Innovations Co. Ltd.) using 10 Fr duct occluder delivery sheath (Cocoon Vascular Innovations Co. Ltd.) After device deployment, an additional PAVF was apparent on LPA angiography, in addition to mild foaming through the device. As oxygen saturation improved from 78% to 93%, the procedure was termed successful and the patient was discharged without procedural complication. 7 days post procedure patient presented with breathlessness and 3 episodes of hemoptysis for 3 days. On examination chest was bilateral clear, with no murmur. Chest X-ray revealed homogenous opacity in left middle zone. His oxygen saturation at room air was 93%. In view of suspected pulmonary infarction patient was managed conservatively with cough suppressants, antibiotics and tranexaminic acid for 7 days. Patient improved on this treatment and was discharged after 10 days. At 1 year follow up patient is doing fine with no recurrent hemoptysis or breathlessness with oxygen saturation of 93%.

This case highlights that there can be multiple PAVF in a patient, whether all should be occluded is debatable. In our patient the small PAVF was not closed and at one year follow up patient is doing well with oxygen saturation of 93%. Retrograde approach to close the PAVF is other option if there are multiple feeders with single draining channel. Pulmonary infarction can occur post device closure of PAVF which can be conservatively managed.
Figure 2 – Duct occlude device in situ with mild foaming through the device.
Figure 1 – Computed Tomography of pulmonary angiography showing a large pulmonary arteriovenous fistula from left pulmonary artery with feeding artery measuring 14.5mm and draining channel measuring 12.5mm.