the false lumen. However, perfusion through a femoral artery (16F) caused right cerebral malperfusion. Surprisingly, addition of right axillary artery perfusion (12F) was ineffective, and malperfusion persisted even after innominate artery occlusion, putting this patient at great risk. Nevertheless, the clinical outcome was not affected because right cerebral blood flow was restored several minutes later, although the exact mechanism of that remains unknown. Moreover, tight snaring or clamping of a dissected artery may cause another vascular injury and is not an optimal maneuver [1].

Our patient had a small build, and the size of the arterial cannula was determined in proportion to both arteries. Therefore, the true cause of malperfusion in this case was a pitfall of the CPB strategy. Although right axillary arterial perfusion has recently been advocated during surgery for AAD, and some even claim that a very small cannula is acceptable for the axillary artery [2], that is not always the case. In contrast, axillary artery perfusion is not reliable, and cerebral rSO₂ monitoring is indispensable even in the absence of critical findings, such as seen with the present patient. A dead-end false lumen in the innominate and carotid arteries requires special caution because CPB flow may expand it to cause cerebral malperfusion. Innominate artery occlusion can be an effective emergency measure in some cases in which a dual artery perfusion strategy is adopted.

There is a growing body of evidence showing that perfusion through the axillary artery can cause cerebral malperfusion [1, 3, 4]. Moreover, in patients with carotid artery obstruction, thrombi may exist in the true lumen, and simple flow restoration through the axillary artery can be hazardous [5]. If AAD obviously affects the innominate or carotid artery, direct carotid artery cannulation may be the method of choice.

Fig 2. Immediately after initiation of cardiopulmonary bypass, the regional oxygen saturation of the right cerebral hemisphere (red line) disproportionally fell (x). After cardiopulmonary bypass was restarted using both the right femoral and right axillary arteries for arterial inflow, the regional oxygen saturation of the right side of the brain fell again (y). Distal aortic anastomosis under retrograde cerebral perfusion (z). (Blue line = left.)

References

Unexpected Cause of Cyanosis and Dyspnea in an Adult: Direct Communication of the Right Pulmonary Artery and Left Atrium

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Direct communication of the right pulmonary artery and the left atrium is an extremely rare congenital malformation of the pulmonary vasculature. A 41-year-old woman with a history of cyanosis since childhood presented with mild exertional dyspnea. On physical examination, she had central cyanosis, clubbing of the fingers, and an upright position caused by orthodeoxia. Imaging studies showed a very large aneurysm in the distal right pulmonary artery with a direct communication to the left atrium. The patient underwent successful repair, with resolution of hypoxia and exertional symptoms.

(Direct communication of the right pulmonary artery and left atrium is an extremely rare congenital malformation of the pulmonary vasculature. A 41-year-old woman who was a mother of 2 children with a history of cyanosis since childhood presented with mild exertional dyspnea. On physical examination, she had central cyanosis, clubbing of the fingers, and an upright position caused by orthodeoxia. Imaging studies showed a very large aneurysm in the distal right pulmonary artery with a direct communication to the left atrium. The patient underwent successful repair, with resolution of hypoxia and exertional symptoms.

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exertional dyspnea. She had no history of trauma or cardiac intervention; however, she had been receiving warfarin for recurrent deep vein thrombosis since her second pregnancy. She also had hearing loss in her right ear.

On physical examination, she had central cyanosis and clubbing of the fingers. There were no hemangiomas or telangiectasias in the skin or mucous membranes of the nose and mouth. Cardiac examination was unremarkable, with no murmurs even with provocative maneuvers. Pulse oximetry with the patient in a supine position at rest showed 75% saturation while breathing room air, which was not altered by nasal oxygen. Moreover, her blood oxygen level dropped to 68% within 1 minute after standing; however, no symptoms developed. Hemoglobin level was 14.1 g/dL, hematocrit value was 55.7, ferritin level was 3.2 ng/mL, and other laboratory test results were within normal levels.

Electrocardiography showed a normal sinus rhythm with no signs of chamber enlargement. Chest roentgenography showed a rounded opacity at the right border of the heart (Fig 1A). Echocardiography demonstrated normal left and right ventricular function and normal valvular function. The left atrial diameter was 36 mm. There was no evidence of pulmonary hypertension. Additionally, an aneurysmal sac that was communicating with the adjacent left atrium was noted (Fig 1B). Injection of agitated saline revealed opacification of the left atrium within 2 cardiac cycles, indicating the presence of a right-to-left shunt. Transesophageal echocardiography confirmed these findings. Computed tomography showed a 60 × 37 mm large aneurysmal sac communicating with the left atrium. Bilateral bronchial arteries were hypertrophied. A right aortic arch was also noted. Additionally, the proximal part of the celiac artery was narrowed significantly, suggesting median arcuate ligament syndrome. Magnetic resonance imaging of the upper abdomen showed a wedge-shaped area of infarction in the spleen, in addition to focal lesions, the differential diagnosis of which included hamartoma or hemangioma. Subsequently, catheterization was performed. The left pulmonary artery was normal. There was a prominent aneurysm in the distal right pulmonary artery with a direct communication with the left atrium, and the patient was referred for surgical intervention (Fig 2). She underwent operation under general anesthesia with double-lumen–catheter endobronchial intubation. A right anterolateral thoracotomy was performed. The right superior pulmonary vein was isolated. The right inferior lobe branch of the pulmonary artery was found beneath the pulmonary veins. The pulmonary arteriovenous fistula was found and ligated with polyester tapes on both sides (Fig 3A, 3B). One of the pulmonary veins was draining very close to the aneurysmal sac, so the aneurysmal sac was reduced with a vascular stapler to avoid an obstruction to the pulmonary venous flow. The patient had an uneventful recovery and was discharged on the sixth day after the operation with an oxygen saturation of 95%.

Comment

Direct communications of the right pulmonary artery and left atrium were classified by De Souza de Silva into 3 types based on the pulmonary venous drainage pattern [1]. According to this classification, a normal pulmonary venous drainage pattern is classified as type I. In type II, the right inferior pulmonary vein is absent and the
pulmonary artery and left atrium are connected by a fistulous connection at the normal site of its origin. In type III, all pulmonary veins connect to the aneurysmal sac. Ohara and colleagues [2] later added a fourth type, in which the right inferior pulmonary vein was replaced by 3 small veins connected to the aneurysmal pouch.

Symptoms and the age at presentation depend on the degree of hypoxemia and the magnitude of the right-to-left shunt and vary from vague symptoms to death in the neonatal period. In severe cases, urgent surgical intervention may be needed, whereas older patients have a milder form of disease and may be asymptomatic or may have exertional dyspnea only [3]. Venous blood bypasses the pulmonary circulation, and bacteria and thrombi in the bloodstream are not filtered in the pulmonary circulation, where they would be removed, which may result in cerebral or systemic embolization or abscess. In addition, hypoxemia causes secondary polycythemia and increases blood viscosity, which also provides an adequate environment for abscess formation. Other possible presentations include congestive heart failure, infective endarteritis, or death related to aneurysm rupture.

Cyanosis and clubbing can be detected during physical examination. Usually an audible murmur does not exist. Unlike pulmonary arteriovenous malformations, telangiectasia is not expected. Aneurysmal sac or possible chamber enlargements can be seen on echocardiography, and Doppler examination may show turbulent flow into the left atrium. If present, additional lesions, mostly atrial septal defects and patent ductus arteriosus, can be established. Contrast echocardiography is very useful in the diagnosis, and bubbles would appear in the left atrium in 2 to 3 cardiac cycles, similar to pulmonary arteriovenous fistulas. Contrast-enhanced computed tomography can delineate the vascular anatomy; however, selective pulmonary angiography remains the gold standard.

The aim of the treatment is to prevent chronic arterial hypoxemia, thromboembolic complications, and heart failure [4]. Surgical management includes ligation, local excision, segmentectomty, lobectomy, or pneumonectomy [5]. Successful percutaneous closure has also been described using coils or Amplatzer devices. However, anatomy of the sac and pulmonary artery, feasibility of the intervention, and associated abnormalities should be considered during the decision making.

Our patient presented with mild exertional dyspnea and cyanosis, and her evaluation revealed a very rare cause of a right-to-left shunt. Such late presentation may result from the vague symptoms, adaptation to mild symptoms, or increase in the size of the aneurysm and shunt magnitude over time [6]. Her hemoglobin levels were lower than expected, probably because of the iron deficiency. Further decrease in arterial blood oxygen saturation when the patient is standing and improvement when supine—orthodeoxia—was also established in this patient. This can be explained by the effect of gravity, causing increased blood flow to the aneurysm located in the base of the right lung. We referred our patient for surgical intervention because of the large aneurysmal sac, which would turn into dead space and could cause systemic embolization if treated percutaneously. Moreover, embolization of the device was a possibility, with catastrophic consequences. This case reminds the clinician of the need to search for pulmonary shunts in patients with cyanosis and without structural heart disease.

References