

Fatal thromboembolism of major aortopulmonary collateral arteries

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Pulmonary atresia with ventricular septal defect (PAVSD) is a complex congenital heart disease, in which pulmonary blood flow depends on major aortopulmonary collateral arteries (MAPCAs). The surgical treatment of PAVSD with hypoplastic native pulmonary arteries consists in intracardiac correction and unifocalization, which provides blood supply from the right ventricle to pulmonary circulation. Although stenosis of MAPCAs and different distribution of blood flow have been observed, there is no evidence on thrombosis in such patients. To our knowledge, this is the first report describing collateral artery thromboembolism in a patient with PAVSD.

A 20-month-old female child with PAVSD, hypoplastic pulmonary arteries, and MAPCA-dependent pulmonary blood flow (FIGURE 1A) was admitted to the Department of Cardiology after hemoptysis and syncope. Similar symptoms occurred several times before admission and were associated with upper respiratory tract infections. On admission, the patient was in poor general condition, with severe cyanosis (pulse oximetry, 50%) and symptoms of right heart failure. Pulmonary embolism was suspected due to elevated D-dimer levels, polycythemia, and thrombocytopenia. Computed tomography angiography confirmed thrombosis of the collateral arteries in the right lung (FIGURE 1B). The effectiveness of treatment with unfractionated heparin was confirmed by computed tomography. However, any changes in anticoagulant therapy were associated with new episodes of severe desaturation. Diffused thrombosis in pulmonary vessels in the right and left lungs was confirmed by cardiac catheterization. The right lower collateral artery was completely occluded (FIGURE 1C), and the right upper collateral artery was

critically stenosed. Alteplase was injected locally to collateral arteries and then continued as an infusion for 72 hours. During control catheterization, balloon angioplasty of the stenosed MAPCA was performed. All these steps did not result in a significant improvement. Alteplase was followed by low-molecular-weight heparin (LMWH) and aspirin. Other supportive treatment, that is, milrinone, dopamine, sildenafil, digoxin, and supplementary oxygen, was provided to increase pulmonary flow. Due to persistent changes in pulmonary vasculature, the patient was considered ineligible for correction with unifocalization, either at our institution or in a foreign cardiac surgery center with experience in unifocalization.

At 1-year follow-up, the patient remained stable on LMWH treatment, with a blood saturation of 70% to 80%. There were no episodes of thrombosis. Thrombophilia was excluded. Angiography showed better perfusion of the lung (FIGURE 1D), but the patient was still ineligible for cardiac surgery.

Regardless of the diversity in pulmonary vascular lesions, MAPCA thrombosis should be considered in the management of patients with PAVSD. Such a diagnosis may influence the choice of treatment and worsen prognosis.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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FIGURE 1

A – aortography showing hypoplastic pulmonary arteries (arrow), major aortopulmonary collateral artery (MAPCA)–dependent pulmonary blood flow; **B** – computed tomography angiography showing thrombosis of MAPCAs in the right lung (arrow); **C** – aortography during MAPCA embolization, showing the right lower collateral artery with complete occlusion (arrow); **D** – aortography at 1-year follow-up showing the distal segment of embolized MAPCA filled by the collateral circulation (arrow)

