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May 2024 Medical Image of the Month: Hereditary Hemorrhagic Telangiectasia in a Patient on Veno-Arterial Extra-Corporeal Membrane Oxygenation

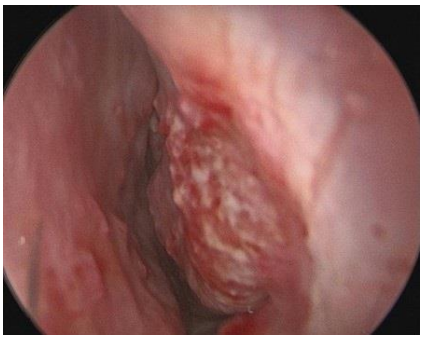

Figure 1. Preoperative nasopharyngoscopic direct visualization of telangiectasia of the nasal turbinate.

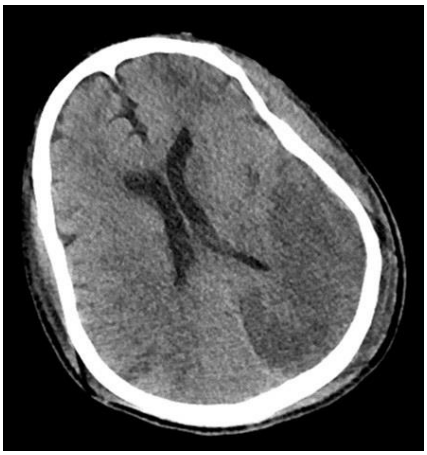

Figure 2. Noncontrast head CT on postoperative day 3 demonstrates extensive multifocal areas of low attenuation consistent with early signs of infarction involving much of the cerebral hemispheres, most prominently involving the left parietal lobe.

A 54-year-old man with a complex cardiac history, including Tetralogy of Fallot requiring Blalock-Taussig shunt in infancy, infundibular patch repair at age 7, and bioprosthetic tricuspid valve replacement at age 52, had ongoing frequent hospitalizations with decompensated right ventricular heart failure secondary to native pulmonary valve mixed stenosis plus regurgitation and left pulmonary artery stenosis. His case was further complicated by his history of hereditary hemorrhagic telangiectasia (HHT) with recurrent epistaxis and recent GI bleeds with multiple angiodysplastic lesions throughout the stomach, duodenum, and descending colon which were previously treated with argon plasma coagulation. The patient was admitted to our hospital in NYHA class IV heart failure receiving a continuous dopamine infusion and aggressive diuresis. Upon admission, a right heart catheterization demonstrated severe pulmonary valve regurgitation, left pulmonary artery stenosis, and systemic hypoxemia suggestive of an intrapulmonary shunt. Admission transthoracic echocardiogram demonstrated normal left ventricular ejection fraction of 55-60%, a severely enlarged right ventricle, moderately reduced right ventricular systolic function, severe pulmonary valve regurgitation, and moderate pulmonary valve stenosis.

A multidisciplinary team including congenital cardiology, pulmonary hypertension, interventional pediatric cardiology, and congenital cardiovascular surgery was consulted and after extensive discussions the patient consented to surgical intervention. Prior to his operative date, he underwent cauterization of his bilateral nasal cavity telangiectasias by Otolaryngology (Figure 1). On hospital day sixteen, he underwent a fourth time redo median sternotomy, pulmonary valve replacement with St. Jude Epic 27-mm porcine bioprosthesis, and repair of left pulmonary artery stenosis. Intraoperative transesophageal echocardiogram at the end of the surgical case demonstrated severe right ventricular dilation, severe right ventricular systolic dysfunction, normal pulmonary valve prosthesis, and left ventricular ejection fraction of 55%. The case was technically challenging requiring a cardiopulmonary bypass time of 178 minutes, and massive transfusion (including twelve units packed red blood cells, two packs of platelets, 4 units fresh frozen plasma, and 10 units cryoprecipitate) for a total estimated blood loss of 3.9 L.

Postoperatively, he had persistent right ventricular systolic dysfunction and diffuse mediastinal hemorrhage. By postoperative day two, a repeat transesophageal echocardiogram revealed worsening right ventricular dilation and severe right ventricle systolic dysfunction. The multidisciplinary care team recommended central venoarterial extracorporeal membrane oxygenation (VA ECMO) support for both worsening hypoxemia and continuing severe right ventricular failure. The aorta was cannulated with a 22 French Bio-Medicus cannula (Medtronic, Minneapolis, USA) and the right atrium cannulated with a 36 French venous cannula (Medtronic, Minneapolis, USA), and full ECMO support was initiated using a Cardiohelp console with a HLS 7.0 oxygenator (Getinge, Goteborg, Sweden) reaching ECMO blood flows of 6 L/minute (an indexed ECMO blood flow of 2.6 L/minute/m^2).

On POD 3, bronchoscopy was performed and revealed diffuse thin bloody secretions in the distal airways without a focal source, which was cleared with suction but quickly reaccumulated. Due to the pulmonary hemorrhage and recent mediastinal hemorrhage, systemic anticoagulation was not started at that time. Due to a lack of awakening during a sedation vacation,

computed tomography (CT) imaging of his head was obtained and demonstrated a large ischemic stroke affecting the majority of the left MCA territory and part of the right parietal lobe (Figure 2).

HHT (also known as Osler-Weber-Rendu disease) is an autosomal dominant genetic disease with various vascular manifestations (1). In addition to the more common mucocutaneous and gastrointestinal tract telangiectasias, some patients with HHT also have pulmonary arteriovenous malformations (AVMs) with right-to-left shunt that can cause hypoxemia with resultant polycythemia. Cerebral AVMs present a risk of intracranial hemorrhage, ischemia, and hydrocephalus, which correlate with the size of the vascular defect. Given the presence of AVMs and hemorrhagic complications related to telangiectasias, the use of extracorporeal membrane oxygenation (ECMO) in patients with HHT is a potentially high-risk situation. This case highlights the risks of ECMO in patients with HHT. The causes of this patient's hemorrhagic and thrombotic events were most likely multifactorial, including contributions from a dilutional and consumptive coagulopathy after cardiopulmonary bypass and hemorrhage, initiation of ECMO, kidney failure, and his underlying HHT. The timing and precise cause of our patient's cerebral infarction are unclear. However, patients with HHT and clinically significant intrapulmonary AVMs may have an increased risk of paradoxical thromboembolic stroke (2). The international HHT expert guidelines assert that even though HHT is a hemorrhagic disorder, it provides no protection against thrombosis (3). In addition, patients with HHT may levels of von Willebrand factor and factor VIII, which would potentially increase their risk of thrombosis (4). This case exemplifies the substantial risks of hemorrhagic and thrombotic complications associated with ECMO for patients with HHT. Further study is needed to help determine whether HHT should be considered a contraindication to ECMO.

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