Incidental Pulmonary Embolus in a Teenager with Gaucher’s Disease Presenting for Interval Bronchoscopy

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Introduction
The occurrence of venous thromboembolism (VTE) and associated sequelae is increasing in pediatrics with improved treatment strategies for childhood diseases and technology for detection. We report a case of a teenager with an uncommon disease who presented for a routine procedure who was found to have an incidental pulmonary embolism (PE).

Case Report
A 17 yo male with Gaucher’s disease presented for surveillance CT scan and then bronchoscopy under general anesthesia. The patient had severe Gaucher’s with history of poor functional capacity and femoral neck infarctions requiring emergency surgery. He was treated previously for PE thought to be due to central venous line (workup otherwise negative). He then had a recurrent PE without known cause but was non-compliant with outpatient anticoagulation therapy. He was on eliglustat trial for Gaucher’s disease with marked improvement in pulmonary symptoms. The day of the procedure he was asymptomatic and preop oxygen saturation was 95%. He had a cast on his foot from recent metatarsal fracture from wakeboarding. During anesthesia induction, the proceduralist was informed that a new saddle PE was present on CT by radiology. This information was relayed to the anesthesia care team during the procedural time out. Bronchoscopy was not performed, anesthesia was completed, and the patient was hospitalized for anticoagulation therapy. No definitive cause was found for his PE; upper and lower extremity DVT scans were negative.

Discussion
Our patient presented a clinical conundrum. He had a rare disease characterized by increased risk of bleeding but had a history of pulmonary embolism. In addition, although asymptomatic, a saddle PE was found on CT.

Gaucher’s disease is the most common of the lysosomal storage diseases. It leads to multisystemic accumulation of glucocerebroside, compromising the function of the spleen, liver, bone marrow, bone mineral, and the lungs. Patients can be prone to bleeding. Anesthetic care may be required for surgery to correct the sequelae of the disease (e.g. partial splenectomy or fixation of pathologic fractures) or during various diagnostic procedures.1

Risk factors of VTE are summarized by Virchow’s triad: stasis, endothelial injury, and hypercoagulability. In children, the most common risk factors are presence of central venous catheter, age (neonates and adolescents), infection, inherited hypercoagulability, trauma, obesity, malignancy, and chronic inflammatory diseases.2

PE typically presents as dyspnea, anxiety, and tachypnea in awake patients and hypotension, tachycardia, hypoxemia, and decreased end-tidal CO2 under GA. The presence of shock and right ventricular failure (right heart strain on CT) are associated with adverse outcomes. The clinical manifestations of PE in children may be non-specific, mimicking the clinical symptoms of the underlying disease. 3 Incidental PE is a challenge in clinical practice. Current management strategies are the same for symptomatic PE.4

Combined CT/bronchoscopy cases have become common to minimize anesthetic exposure. This case emphasizes the importance of team communication and procedural time-outs that should probably include the finding of previous procedures that may not always be conveyed to the patient care team especially on busy clinical days. As in this case, findings may be clinically significant.

Old thrombus in right main pulmonary artery and new thrombus in the left main pulmonary artery extending into all left lobar arteries. The main pulmonary artery is distended and of greater diameter than the aorta. No right heart strain is noted.

References