Spontaneous Paraesophageal Hematoma

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Abstract

Spontaneous paraesophageal hematoma (SPH) likely shares a common etiology with spontaneous intramural esophageal hematoma (IEH). Patients with IEH typically present with hematemesis or melena leading to early detection and management, but patients with SPH do not have overt gastrointestinal bleeding on presentation. Management depends on the correction of the underlying causative factor. We present the first case of a spontaneous paraesophageal hematoma in a patient with hemophilia B. Awareness of this complication of hemophilia, its clinical manifestations, and imaging findings, allows for a timely diagnosis and appropriate management.

Introduction

Gastrointestinal (GI) bleeding is a potentially disastrous complication occurring in 25% of patients with hemophilia A and B (deficiency of coagulation factor VIII and factor IX, respectively).¹,² Typically, GI bleeding is precipitated by minor or major trauma, and patients present with overt hematemesis or melena. Intramural hematomas usually present after mechanical trauma secondary to emesis, which leads to increase in esophageal pressure. Spontaneous intramural hemorrhage of the GI tract occurring in the esophagus, stomach, small intestine, and colon has been described in patients with hemophilia or in patients taking anticoagulants, and etiology of such hemorrhage is thought to be secondary to minor trauma to the mucosa.³,⁴ Spontaneous paraesophageal hematoma (SPH) has yet to be described in a patient with hemophilia.

Case Report

A 24-year-old man with severe hemophilia B (factor IX level less than 1%) presented with sharp, non-radiating, acute retrosternal chest pain for 1 day that was worsened by oral intake. There were no precipitating or alleviating factors. He reported a history gastrointestinal esophageal reflux but denied emesis, trauma, NSAID or alcohol use, or GI bleeding. Previous hemophilia-related complications included hemarthrosis and intramuscular bleeding. His only medications were 4,000 units of coagulation factor IX as needed. On examination, his vital signs were stable and there were no ecchymosis or signs of trauma to the chest wall. Physical examination was unremarkable except for a right wrist ecchymosis, which was attributed to minor trauma sustained at work. His hemoglobin was 13 gm/dL, and his activated partial thromboplastin time was 126, consistent with history of hemophilia B. Chest computed tomography (CT) showed a paraesophageal hematoma measuring 3 x 1.8 cm surrounding the distal 8.5 cm of the esophagus with mural thickening and inflammatory changes (Figure 1). He was managed conservatively and treated with antacids, clear liquid diet, and 50 units/kg of coagulation factor IX every 12 hours for 5 doses. His factor IX level was maintained at >50%. An esophagogram did not reveal any perforation, and his symptoms resolved after 4 days. He was discharged and remained asymptomatic on outpatient follow-up.
Spontaneous IEH in patients with hemophilia is an uncommon entity, presumably caused by mucosal injury of the GI tract. In patients with coagulopathy, intramural hematomas of the esophagus typically occur after injury like vomiting, pill-induced esophageal injury, foreign body ingestion, or endoscopic procedures. Presenting symptoms include chest pain, dysphagia, hematemesis, or melena. SPH may also present with retrosternal chest pain, but because paraesophageal veins are located adjacent to the esophageal wall, an inciting factor such as trauma or vomiting is not necessary to cause a spontaneous bleed. Thus, hematemesis or other overt clinical signs of GI bleeding frequently seen in IEH are not usually present in SPH.

Early imaging studies are required to diagnose and treat such complications. We suspect that our patient’s rare paraesophageal hematoma resulted from spontaneous bleeding of a paraesophageal vein triggered by his underlying coagulopathy. The atypical nature of his bleeding episode may be explained by the severity of his disease and by not administering prophylactic factor IX. Treatment with replacement factor IX to correct the coagulopathy and conservative management led to symptomatic improvement and resolution of the hematoma. It is crucial to consider spontaneous paraesophageal hematomas in patients with hemophilia who present with atypical chest pain and do not have clinical evidence of GI bleeding. A timely diagnosis may prevent enlarging hematoma formation, esophageal compression, and potentially life-threatening rupture. Early imaging and detection can give an opportunity to correct the underlying causative factor, leading to excellent clinical outcomes.

Disclosures
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Figure 1. Axial contrast-enhanced CT showing (A) a paraesophageal collection and punctate hyperdensity concerning for site of bleeding (arrow), and (B) a paraesophageal hematoma (*) adjacent to a thickened esophagus (arrowheads). Note enhancement along the esophageal adventitia and the adjacent collection, consistent with paraesophageal collection. (C) Coronal contrast-enhanced CT showing collection adjacent to esophagus (*) and a punctate focus of hyperdensity concerning for a site of active bleeding (arrow).