Primary small-bowel diffuse large B-cell lymphoma presenting as hematemesis

A 32-year-old man with a past medical history of gastroesophageal reflux disease and gastric ulcers presented to the emergency department with hematemesis. He endorsed epigastric pain, one melanotic bowel movement, dizziness, and weakness. He denied previous episodes, aspirin or nonsteroidal anti-inflammatory drug use, and alcohol consumption.

On examination, the patient had hypotension and tachycardia, with epigastric tenderness in response to palpation and black stool on rectal examination. Laboratory test results indicated microcytic anemia, with a hemoglobin level of 8.0 g/dL that dropped to 6.2 g/dL after fluid resuscitation.

The patient was admitted to the intensive care unit, received blood products and intravenous esomeprazole, and underwent emergency esophagogastroduodenoscopy (EGD). This revealed blood in the gastric chamber and an ulcerated, oozing, circumferentially infiltrative mucosa in the second portion of the duodenum (Fig. 1a, Fig. 1b, Fig. 1c). Abdominal computed tomography displayed localized thickening of the duodenal wall (Fig. 2).

During a second EGD, oozing from the lesion had ceased, so that tissue sampling was possible (Fig. 1d). The histological and immunohistochemical findings were consistent with diffuse large B-cell lymphoma (DLBCL) (Fig. 3a, Fig. 3b, Fig. 3c, Fig. 3d, Fig. 3e, Fig. 3f). Positron emission tomography (PET) revealed an area of increased metabolic activity in the second portion of the duodenum (Fig. 4a, Fig. 4b).

The patient’s treatment consisted of six cycles of chemotherapy with rituximab (Rituxan, Genentech and Biogen Idec), cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) given every 3 weeks. The patient tolerated the chemotherapy adequately, and interim PET after three cycles revealed a complete response to treatment (Fig. 4c, Fig. 4d). The patient’s clinical course and imaging are summarized in Video 1.

The gastrointestinal tract is the most common site of extranodal non-Hodgkin lymphoma. However, isolated primary lesions of the duodenum are rare, accounting for fewer than 0.8% of the cases [1]. The manifestations of gastrointestinal lymphoma vary with the subtype and...
anatomical location. For DLBCL, these frequently include weight loss, obstructive symptoms, abdominal pain, and lower gastrointestinal bleeding [2, 3]. This case is important in that it is the first well-documented case of intestinal DLBCL presenting with hematemesis.

Fig. 3  Histology. a The duodenal mucosa shows an underlying cellular infiltrate within the lamina propria (hematoxylin and eosin [H&E] stain, magnification × 100). b The cellular infiltrate comprises large atypical lymphocytes with scant cytoplasm, marked nuclear enlargement, variably prominent nucleoli, and increased mitotic figures (H&E stain, magnification × 400). c–f The atypical lymphocytes are immunoreactive for pan-B cell marker CD20. c Magnification × 200, co-expression of BCL-2. d Magnification × 200, co-expression of MUM-1. e Magnification × 200, Ki-67 labeling index approximately 60%. f Magnification × 100.
Competing interests: None

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Acknowledgments

We would like to thank Dr. Sarayu Chandrashekhar for her invaluable assistance in the pathologic interpretation of the tissue samples.

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DOI http://dx.doi.org/10.1055/s-0034-1392874
Endoscopy 2015; 47: E526 – E528
© Georg Thieme Verlag KG Stuttgart · New York
ISSN 0013-726X

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Fig. 4  Positron emission tomography. a, b Before treatment. Diffuse concentric mural thickening involving a short duodenal segment measuring 2.7 cm. Robust metabolic activity and a maximum standardized uptake value (SUV) of 14.6. No additional anatomical sites of extranodal, splenic, or nodal involvement are identified in the rest of the body. c, d After three cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP). Complete response to treatment is seen on the interim axial and coronal images, as evidenced by a lack of hypermetabolic activity in the previously affected area corresponding to the duodenum and minimal residual nonconcentric mural thickening.