A very rare case of extraskeletal Ewing sarcoma of the duodenum presenting as gastrointestinal hemorrhage

A 51-year-old man presented to our hospital with hematochezia, melena, palpitations, and lightheadedness for 1 week. The patient was found to have anemia with a hemoglobin concentration of 73 g/L and a peripheral blood smear suggestive of hypochromic microcytic anemia. To confirm upper gastrointestinal bleeding, an esophagogastroduodenoscopy was performed, revealing a protruding lesion with surface ulceration and active bleeding at the inferior flexure of the duodenum (▶ Fig. 1a). The lesion was located on top of a submucosal elevation of approximately 3×2.5 cm in size (▶ Fig. 1b, c). He received endoscopic hemostasis, including epinephrine injection, argon plasma coagulation, and hemoclipping (▶ Fig. 1d, Video 1). The biopsy was suggestive of small cell carcinoma. To obtain more details for the optimal surgical approach, abdominal computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) showed a target-like lesion with eccentric wall thickening at the inferior flexure of the duodenum, measuring approximately 2.8×2.4 cm in diameter (▶ Fig. 2a–c).

The patient underwent a laparoscopic pancreaticoduodenectomy for a duodenal carcinoma. Gross examination was notable for an ulcerative tumor of approximately 3×3×2.5 cm in diameter at the inferior flexure of the duodenum, which had gray-white cup-like shape appearance with solid and cauliflower-like areas (▶ Fig. 2d, e; Video 1). Histological and immunohistochemical examination revealed poorly differentiated extraskeletal Ewing sarcoma of the duodenum, which was positive for CD99/FLI-1/INI-1/Ki67(60%)/NKX2.2 but negative for CK-P/LCA/S100/CD56/WT-1/desmin (▶ Fig. 3). Upon genetic testing, a fusion of the EWSR1 gene on chromosome 22q12 was found with the gene encoding the transcription factor ERG (exon 8 of EWSR1 fused to exon 9 of ERG and a wild-type KRAS/NRAS/BRAF. Postoperatively, the patient was treated with interval-compressed VDC/IE chemotherapy. Two months after surgery, the CT showed no sign of tumor recurrence or enlarged lymph nodes (▶ Fig. 2f).

Extraskeletal Ewing sarcoma of the duodenum is an extremely rare tumor with a poor prognosis [1]. The diagnosis of extraskeletal Ewing sarcoma of the duodenum is complicated because of the lack of specific clinical symptoms and imaging findings [2]. There is no treatment guideline for this type of cancer. Our case suggests that patients presenting with recurrent hematochezia, weight loss, an abdominal mass, and anemia should receive esophagogastroduodenoscopy for diagnosis. It seems that surgical resection and adjuvant chemotherapy may contribute to a relatively good survival outcome for patients with extraskeletal Ewing sarcoma of the duodenum.

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Competing interests

The authors declare that they have no conflict of interest.
The authors

Zilong Zhang‡, Xin He‡, Songqi Wen, Xin Jin, Ding Xiao, Jian You
Department of Hepatobiliary-Pancreatic and Hernia Surgery, Wuhan Fourth Hospital, Puai Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan, China

Corresponding author

Jian You, MD
Department of Hepatobiliary-Pancreatic and Hernia Surgery, Wuhan Fourth Hospital, Puai Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430033, China
Fax: +86-28 85423052
jianyou67@163.com

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Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

‡ Contributed equally to this work.
Fig. 3  Tumor histology consistent with poorly differentiated small round cell tumor. 

- **a** Hematoxylin and eosin, × 200, with no lymphovascular invasion and a negative vertical margin. 
- **b** Immunohistochemically, the Ki-67 (× 200) index of small round cell tumor cells was 60%. 
- **c-e** Tumor positive for CD99 (× 200) and FIL-1 (× 200), INI-1 (× 200). 
- **f** Genetic testing showed EWSR1–ERG fusion.