Life-threatening bleeding of a duodenal gastrointestinal stromal tumor in a teenager: a rare case report

Piero V. Valli1, Carlo Valli2, Thomas Pfammatter3, Peter Bauerfeind1

1 Clinic of Gastroenterology and Hepatology, University Hospital Zürich, Switzerland
2 Department of Internal Medicine, Gastroenterology, Spitalzentrum Oberwallis, Visp, Switzerland
3 Institute of Diagnostic and Interventional Radiology, University Hospital Zürich, Switzerland

Introduction

Gastrointestinal stromal tumors (GIST) are rare mesenchymal and typically subepithelial neoplasms that account for approximately 1% of the gastrointestinal tumors [1]. Predilection sites are the stomach and the small intestine [2]. Duodenal GISTs are even more rare and exceptional in children or young adults. GISTs most commonly originate from the interstitial cells of Cajal (ICC) in the fourth layer of the gastrointestinal tract wall (muscularis propria). Diagnosis is confirmed by expression of positive immunohistochemical staining for CD117 (KIT receptor tyrosine kinase, c-KIT protein), which is found in 95% of cases. CD34 stains positive in 70% of GIST. The overall GIST incidence is estimated to range between 10 to 20 cases per million among the adult population [1]. GISTs in childhood either occur sporadically or in the context of hereditary syndromes like neurofibromatosis type 1 (NF1) or Carney-Stratakis syndrome. Nevertheless, the occurrence of sporadic duodenal GISTs in children and young adults is exceedingly low. A literature search revealed that only 2 cases of duodenal GISTs in children have been published. Here we report on the case of a 19-year-old female patient who was admitted in hemorrhagic shock due to arterial bleeding of a duodenal GIST located in immediate proximity to the major duodenal papilla. After several attempts of endoscopic hemostasis failed, the tumor bleeding was controlled with a second coil embolization of the pancreaticoduodenal arcades.

Case report

A 19-year-old otherwise healthy female tourist was admitted to a secondary care hospital after fainting while skiing due to suspected gross blood loss with an initial hemoglobin level of 60 g/L. The patient developed tarry stools during the hospitalization. After volume resuscitation including red blood cell (RBC) transfusions, a tumorous mass with a central bleeding ulceration (bull's eye appearance, Fig. 1) was diagnosed upon emergency endoscopy. The submucosal tumor bulging into the duodenal lumen was found in immediate proximity to the major duodenal papilla (Fig. 2). Blood oozing was noted and primary hemostasis was accomplished by injection of saline-diluted epinephrine and the application of 2 Instinct™ endoscopic hemoclips. Non-contrast-enhanced computed tomography failed to locate the primary tumor and did not reveal any distant metastasis. After overnight observation, the patient was transferred to our tertiary care hospital for further diagnostic work-up. Upon arrival at our institution, endosonography demonstrated a submucosal, hypervascular and hypoechoic tumor. The neoplasm with a central bleeding vessel arose from the muscularis propria (fourth wall layer) and measured 25× 15mm (Fig. 3 and Fig. 4). Our initial differential diagnosis based on clinical presentation and endosonographic imaging consisted of gastric stromal tumor (GIST), neuroendocrine tumor (NET), gangliocytic paraganglioma [5], leiomyoma [6] and solid pseudo-papillary tumor of the pancreas [7]. In addition to endosonography-guided fine-needle aspiration, regular biopsies were harvested and an on-site cytologist ensured attainment of diagnostic tissue. Recurrent tumor bleeding after tissue harvesting was then stopped by application of Hemospray®. After observing a recurrent decrease in hemoglobin levels during the following night, ongoing tu-
mor bleeding was confirmed by upper gastrointestinal endoscopy. Given the lack of further endoscopic hemostasis options, transarterial coil embolization of the tumor-supplying anterior pancreaticoduodenal arcade was performed (Fig. 5 and Fig. 6). Despite the first coil embolization, persistent blood loss was noted overnight in the patient. Intermittent bleeding was confirmed by duodenoscopy and no permanent hemostasis was achieved by Gold Probe™ coagulation. Repeat angiography showed persistent tumor staining through tiny branches of the posterior pancreaticoduodenal arcade. The bleeding was finally halted by coil embolization of the inferior pancreaticoduodenal artery via the superior mesenteric artery and the origin of the posterior arcade via the gastroduodenal artery. The diagnosis of a GIST was ultimately established by positive staining for CD117 (cKit), CD34 and DOG–1 and negative staining for SMA und S100. After no further bleeding was detected over the course of the next 48 hours, the patient was transferred to a center in her home country for further treatment. After an initial trial of tumor downsizing with imatinib, the medication had to be discontinued due to severe adverse events including vomiting, diarrhea, fatigue and severe bone marrow suppression requiring multiple transfusions. Thereafter, the GIST was removed entirely by a pancreas-sparing duodenectomy. The final histology report of the in toto resected GIST (positive staining for CD117, CD34 and DOG–1; negativity for Desmin, S11 and CD31) displayed 3 mitoses/50 HPF. These findings correspond to a low risk (8.3%) for progressive disease [8]. The further postoperative recovery of the patient has been uneventful so far.

Discussion

We here report the third case in literature of a duodenal GIST in a patient younger than age 20. The initial presentation of the tumor in our teenage patient was a life-threatening arterial tumor bleeding with consecutive hemorrhagic shock. Even though pediatric GISTs frequently present with gastrointestinal bleeding...
creatic duct, OTSC® application on the juxtapapillary lesion was failed to adequately control the life-threatening tumor bleeding. Fearing a complete closure of the common biliary and the gastrointestinal blood loss, iron deficiency anemia and unexplained gastrointestinal pain. Given the fairly high incidence of metastasis (approx. 50%) and the elevated recurrence rate, early diagnosis of GIST is even more crucial in young patients with a potentially long life expectancy [12].

Competing interests: None

References

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