Introduction

Primary duodenal adenocarcinoma (PDA) is an uncommon malignant neoplasm with a prevalence of only 0.35% among all gastrointestinal carcinomas and 30% to 45% among small-intestinal cancers (1-3). PDA was first described by Hamburg in 1746, and since then most of the reports have included only small numbers of patients (4-7). During the past two decades, the development of endoscopic and radiologic diagnostic imaging and their widespread use have increased the number of PDAs diagnosed.

The low incidence of duodenal carcinoma may be due to several theoretical factors including small bowel transit time, host immunologic factors, and or epithelial toxin exposure. These malignancies are usually diagnosed later and the most common symptom of the third and fourth portions of duodenum is upper abdominal pain related to partial duodenal obstruction and gastrointestinal bleeding (8,9). The mean age of presentation of PDA is 64 with a range of 47-87 years.

Segmental resection is the treatment of choice but it is not clear from literature. We report the case of a patient with adenocarcinoma of angle of Treitz who presented obstruction and underwent segmental resection of duodeno-jejunal junction.

Case report

A 70 year old Italian male was referred to our hospital for intermittent abdominal pain, melena, nausea and vomiting progressive asthenia and anemia. His medical history was significant for type 2 diabetes mellitus, hypertension, coronary heart disease and moderate BPCO. On admission in our department the patient was in mediocre general and nutritional conditions, his blood pressure was 100/60 mmHg, temperature 37°C. Physical exam was unremarkable with the exception of pain in the epigastric and left upper quadrant of abdomen.

Laboratory tests gave the following result: WBC 10,600/mm³ with 52% neutrophils and 26.9% lymphocytes, RBS 3,450,000/mm³.
Committee on Cancer [AJCC] classification) of the duodenum was made. The postoperative course was uneventful. Patient was given chemotherapy and at six months of follow-up is doing well.

On exploration there was a 3.5 cm growth at duodeno-jejunal junction with enlarged mesenteric lymph nodes, without involvement of adjacent structures. Segmental resection of duodenojejunal (D-J) junction with growth with 1.5 cm of margin and clearance of loco regional lymphnodes was done. Histopathology confirmed a poor differentiated adenocarcinoma, pT3N2G3 (American Joint Committee on Cancer [AJCC] classification) of the duodenum was made. The postoperative course was uneventful. Patient was given chemotherapy and chemotherapy and at six months of follow-up is doing well.

**Discussion**

Neoplasms of the small bowel are rare, despite the fact that the small bowel constitutes over 60% of the intestinal tract. Ninety percent of small bowel neoplasms are benign lesions, the majority of which are leiomyomas (41%). Malignant lesions of the duodenum include adenocarcinomas, sarcomas, neuroendocrine tumors, lymphomas, and metastatic lesions. The dominant malignant lesion is adenocarcinoma, which accounts for 37% to 40% of small bowel malignancies (10). The reasons for the low incidence of carcinoma of the midgut are not well understood, but are felt to be related to a short transit time of potentially carcinogenic luminal contents, low bacterial flora load, an alkaline environment, and a low rate of activated precarcinogenic enzymes (11). Adenocarcinomas of the IV portion of duodenum, most commonly present with patients complaining of vague abdominal pain syndromes (12,13). Other symptoms include nausea and vomiting, weight loss, gastrointestinal bleeding, obstruction, and fatigue caused by anemia (12-14). The majority of patients are men in most series (52-67%) (13,15). The mean time from onset of symptoms to diagnosis approximately 4 months (range 1 month to 1.2 years) (12). Diagnosis of adenocarcinoma of the duodenum remains challenging, as direct visual endoscopic access to the midgut lumen is limited. The majority of lesions are diagnosed by radiographic imaging. Diagnostic radiographic studies include ultrasound, computed tomography (CT), and capsule endoscopy. In the Anderson case series (14) an upper intestinal barium study was the most frequent radiographic diagnostic tool to demonstrate a probable neoplasm (diagnostic in 22% of cases), but his series predates the use of capsule endoscopy and modern helical CT imaging (14). CT enteroloscopy utilizes spiral and multidetector row CT technology with a volume challenge of 2 liters of enteral contrast agent administered through a nasojejunal catheter. This technique has become the radiographic diagnostic tool choice for suspected small bowel neoplasm, with a sensitivity an specificity rate of 100% and 95%, respectively (16-17, 18). Occhi (18) et al found that upper GI endoscopy had a 36% false negative result rate in identifying duodenal tumors. Push enteroscopy (PE) provides many benefits including direct visualization of lesions in the proximal duodenum and jejunum, allowing the ability to biopsy and provide therapeutic measures in cases of bleeding. The investigation of obscure bleeding by PE may find a diagnostic cause in 25-28% of cases (19-20). Duodenal first and second part tumors are treated by Whipple’s procedure. For resectable cancers of third and fourth part of duodenum, segmental resection is the treatment of choice. Duodenojejunal segmentectomy is the treatment of choice for Treitz tumors with lymph node clearance; lymph node positivity does not preclude resection (21). The prognosis of these tumors is good (22).

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The mean overall survival is 13 months, with a 30% 5-year survival rate. Factors that reduce long-term survival probability include metastases at diagnosis, unresectability, positive regional lymph nodes, and poorly differentiated tumors (12,14). Cunningham and colleagues reported a 23- month median survival in patients who underwent complete resection of a small bowel adenocarcinoma versus only a 7-month median survival in pa-
Patients undergoing incomplete or palliative surgery (14). The role of adjuvant chemotherapy and radiotherapy in small bowel adenocarcinoma is not clearly defined. Some studies showed that adjuvant treatment after complete resection does not diminish the risk of subsequent recurrence (8).

Conclusion

Adenocarcinoma of the duodenum remains a rare disease, though the prevalence appears to be rising, possibly due to improved diagnostic techniques. Surgery remains the primary treatment for small bowel adenocarcinomas, with a curative resection of locally contained disease significantly improving overall survival. Long-term survival for patients with duodenal adenocarcinoma can be achieved by surgery obtaining negative resection margins. Whipple procedure is required to achieve this goal for most lesions in the first and second part of duodenum. Segmental resection can be appropriate in select patients, especially with lesions of the distal duodenum.

References