Case Report

Integrative Gastroenterology and Hepatology

Diagnostic and Management Difficulties in the Adenocarcinoma Third Part of Duodenum: A Case Discussion

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Abstract

Duodenal Adenocarcinomas (DA) represent approximately 0.3% of all malignant gastrointestinal tumors. DA is the most common site for adenocarcinoma in gastrointestinal track. The most common site of DA within duodenum is the 2nd part of duodenum. A high degree of suspicion is required for the diagnosis of third/fourth part DA. Push enteroscopy is the definitive diagnostic tool for third/fourth part DA. We discussed a case of adenocarcinomas third part of the duodenum with brief discussion on controversies in optimal surgical management.

Keywords: Adenocarcinoma third part duodenum, Difficult endoscopy, Pancreaticoduodenectomy, Pancreas-sparing duodenectomy, Segmental duodenectomy, Push endoscopy, Whipple's operation

Introduction

Bowel obstruction involving third part of duodenum (D3) is rarely encountered in clinical practice. In children benign pathologies such as malrotation, duodenal web, duodenal perforated diaphragm, internal paraduodenal hernia, and tropical jejunoileitis are the usual causes of D3 obstruction [1]. In adults, benign causes of D3 obstruction are superior mesenteric artery syndrome, chronic duodenal ileus, suprarenal aortic aneurysm, tuberculous stricture, and benign idiopathic stricture [2]. The malignant causes of adult D3 obstruction are adenocarcinoma, carcinoid, lymphoma, leiomyosarcoma and gastrointestinal stromal tumor [3]. Adenocarcinoma of small bowel accounts for less than one per cent of all gastrointestinal malignancies [3],[4],[5]. Around half of all the small bowel adenocarcinomas are located in the duodenum [6],[7]. Among the four segments of duodenum the ampullary region of second part of

duodenum (D2) is the most common site of involvement [6]. We herein report a case of duodenal adenocarcinoma (DA) involving D3 with brief discussion on diagnostic difficulties and the management controversies of this aggressive malignancy.

Case Report

A 65-year-old woman presented with intermittent bilious vomiting, post prandial abdominal pain and weight loss of more than 10 kgs for two months. There was no history of fever, jaundice, melena, haematemesis, dyspepsia, corrosive ingestion or prolonged intake of any medication. She was neither addicted to alcohol nor to smoking. Clinical examination of abdomen was normal. Systematic examination was non contributory except for pallor. She was found to have microcytic hypochromic anemia with a hemoglobin level of 8.0 gm/dl. Conventional oesophago-gastro-duodenoscopy

(OGD) documented minimal dilatation of first part of duodenum (D1) and (D2). Advanced age of the patient, acute history and abnormal D2/D1 dilatation was highly suspicious for gastrointestinal malignancy. Elevated levels of gastrointestinal and pancreatic malignancy tumor markers like Carcinoembryonic Antigen (CEA), carbohydrate antigen 19-9 (CA 19-9), and CA 125 further strengthened the possibility of a malignant lesion.

Contrast-enhanced Computed Tomography (CECT) abdomen revealed dilated stomach and D2 without any associated periportal/perigastric/peripancreatic lymphadenopathy. The Portal vein, celiac trunk, superior mesenteric vessels, pancreas and extra hepatic biliary tract were reported to be normal. Repeat OGD was carried out using an extra long fiber scope which identified an ulcero-proliferative growth resulting in near complete occlusion of D3. Endoscopic tissue biopsy demonstrated presence of moderately differentiated adenocarcinoma of duodenum.

The patient underwent exploratory laparotomy under general anesthesia. A solid mass of 2.5×2 cm was identified at the D3 just to the right side of superior mesenteric vessels. Pancreaticoduodenectomy was contemplated after ascertaining the respectability of the neoplasm. We opted for pylorus preserving pancreaticoduodenectomy instead of segmental resection due to lack of frozen section facility at our center. End to side pancreaticojejunostomy with duct to mucosa anastomosis over a stent followed by end-to-side stent less, single layer, interrupted hepaticojejunostomy to a single retrocolic jejunal loop was performed. The bowel continuity was restored by infracolic roux-en-y gastrojejunostomy and enteroenterostomy. Additionally aortocaval lymph node sampling and a feeding jejunostomy was carried out (Figure 1). Histopathological examination of the specimen revealed a moderately differentiated signet variety adenocarcinoma, measuring approximately 3.1×2 cm infiltrating the serosa of duodenal wall, with lymphovascular infiltration with metastatic peripancreatic lymph node (5 metastatic/7 total) and spared aorto-caval lymph node (pT_3 p N_2M_0) (Figure 2). Stage III B as per American Joint Committee on Cancer (AJCC) 7th edition staging. The specimen's margins were free of tumor (Figure 2).

Broad-spectrum antibiotics, H2 blocker and Octeotride were administered in three daily doses of 100 µg for 7 days in postoperative period. On 4th postoperative day, the patient developed severe acute pancreatitis. Despite of

best possible resuscitative efforts she developed ongoing multiple organ failure and died on 15th postoperative day.

Figure 1: (a) Endoscopic view with extra-long fiber optic scopes of the duodenum showing focal segmental luminal narrowing with a reddish area with mucosal irregularity at third part of duodenum (b). Intraoperative photograph showing superior mesenteric vein after pancreatic head excision. (c). Gastrojejunostomy. (d). En-bloc specimen.

Figure 2: Histopathologic evaluation of the excised specimen verified a moderately differentiated adenocarcinoma, infiltrating the duodenal wall.

Discussion

DA represents approximately 0.3% of all malignant gastrointestinal tumors. Predisposing factors for DA are duodenal adenomas (malignant transformation rate of 30-80 %), Familial adenomatous polyposis, Gardner syndrome, and duodenal polyps [8]. The relative frequency of DA in Goldner and Stabile study was: D2 (87%), D3 (7%), and D4 (6%). There are very few reported cases of D1 adenocarcinoma in English literature [8].

The most common presenting symptom for DA is postprandial abdominal pain [8]. Other clinical

manifestations are nausea, vomiting, weight loss, anemia, fatigue, weakness, and gastrointestinal bleeding. This clinical presentation is a great mimicker of acid peptic diseases [9]. The high intestinal obstruction is a relatively late feature of D3 adenocarcinoma [4],[10],[11]. In our case the only clue to the diagnosis of D3 adenocarcinoma was lack of symptomatic improvement with proton pump inhibitors and minimal dilatation of D1 & D2 on OGD. Goldner and Stabile described four stages of disease severity at the time of diagnosis of DA: Stage 0 (10%), Stage I (10%), Stage II (17%), Stage III (50%), and Stage IV (13%) AJCC 7th edition [7].

The findings in upper gastrointestinal contrast series in D3 malignancy can be short segment stenosis, a filling defect or an ulcer crater [12]. The most common CECT feature of D3 malignancy is an exophytic or intramural mass with central necrosis and ulceration. Entirely intraluminal location indicates benign nature of tumour [9]. Abdominal CECT didn't pick up any mass lesion in our patient the reason may be reduced sensitivity of CECT for tumor < 2 cm in dimension. The D3 is often inaccessible by conventional endoscopy, so push enteroscopy is advocated for diagnostic purpose which can reach up to proximal jejunum (about 50-70 cm behind the pylorus) [13],[14]. Endoscopic ultrasound and magnetic resonance imaging are useful for the diagnosis, staging, and determination of resectability of duodenal tumors [9], but were not advised in the index case as tissue diagnosis and CECT was enough to ascertain the diagnosis and resectability.

For non ampullary DA, curative R0 resection options include local wide excision, duodenal segmentectomy after intestinal derotation or pancreaticoduodenectomy/ Whipples' operation [15]. The endoscopic resection is described for early tumours (Tis and T1) [15]. Due to rarity of D3 adenocarcinoma the choice of procedure is difficult to be ascertained [11],[16],[17]. Regardless of the type of surgery, a curative tumor free resected margin has a significant survival advantage compared to non curative resection or chemo-radiation [11]. Radiotherapy as the sole modality of treatment is not beneficial as DA is radio-resistant and poor tolerance of small bowel to radiation [18].

Vascular encasement, invasion of contiguous organs other than the head of pancreas, distant lymphadenopathy (para-aortic), or unresectable metastases precludes curative resection [1]. Majority of D3 tumours with serosal infiltration have local and distant invasion as encountered

in our case. In advanced and unresectable nonampullary DA palliation can be offered via non surgical means such as stenting via deep enteroscope, or surgical means like duodenojejunostomy or gastrojejunostomy [11], [16],[17].

The surgical complications following pancreaticoduodenectomy are pancreatic, biliary and enteric fistula, pancreatitis, stress ulcer hemorrhage or diffuse retroperitoneal hemorrhage, cholangitis and intra abdominal abscess. Medical complications (4%-19% of patients) can be cardiac, respiratory, renal dysfunction, pulmonary embolism, hepatic and metabolic dysfunction. Our patient died because of acute fulminant pancreatitis leading to fatal systemic inflammatory response syndrome [6].

After curative resection adjuvant chemotherapy should be considered for stage III disease [7],[17],[18]. Tumor stage, resectability and nodal involvement are independent predictors of survival in D3 adenocarcinoma [18]. Five year survival following curative resection is up to 50%, while unresectable disease has a very poor prognosis [18]. Five year survival following R0 resection can reach up to 62% in adenocarcinoma involving distal duodenum and without lymph node metastasis [2]. The node-positive patients have a statistically higher recurrence rate than their node negative counterparts [2].

To conclude D3 adenocarcinoma needs a high index of suspicion for diagnosis. Upper gastrointestinal tract contrast series and OGD seem to be the most accurate diagnostic modalities when evaluating patients with suspicious symptoms and signs for proximal small bowel malignancies. Early diagnosis and curative resection offers a very good prognosis contrary to that in periampullary and supra ampullary adenocarcinoma [19].

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