Case Report

A rare case of duodenal cancer with achalasia cardia

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Abstract

Duodenal adenocarcinoma constitutes 0.4% of gastrointestinal malignancies. Achalasia incidence rate is 0.5-1.2 per 100000. The combination is rare.

This is a report of a 68-year-old male from Nepal with history of five years abdominal pain, dysphasia and weight loss. Duodenoscopy could confirm ulcero-proliferative growth at D1-D2. Barium meal depicted features of achalasia cardia.

No similar case report suggests that occurrence of duodenal carcinoma and achalasia cardia is merely co-incidental.

Keywords: Achalasia cardia; Duodenal cancer; Heller's cardiomyotomy; Whipple's operation.

Introduction

Duodenal adenocarcinoma is a rare malignancy that constitutes 0.4% of gastrointestinal malignancies but 45% of all small bowel adenocarcinomas.[1] Despite advancements in techniques of diagnosis and resection and decreased perioperative mortality and morbidity, 5-year survival is only 45–55%.[1-3]

Achalasia is a neurodegenerative motility disorder of the esophagus resulting in deranged esophageal peristalsis and loss of lower esophageal sphincter function. Historically, annual achalasia incidence rates were believed to be low, approximately 0.5-1.2 per 100000. More recent reports suggest that annual incidence rates have risen to 1.6 per 100000 in some populations.[4] The combination of these two clinical cases is even rarer. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Case report

This is a report of a 68-year-old male from Nepal, who was admitted to the surgical gastroenterology unit with five years history of upper abdominal pain, indigestion, dysphasia and weight loss of greater than 6 kgs within two months. The patient used to be a smoker and an alcoholic but stopped as his symptoms progressed. Despite 10 months of proton pump inhibitor treatment, his complaints were still present. There were no comorbidities.

At presentation his body mass index was 22.76 kg/m^2, Albumin 4.1 gm/l, Nutritional risk index was 103.9, ECOG 1, Karnofsky Performance Score 90. Systemic examination revealed no abnormalities.

Laboratory investigations revealed Hb of 13gm/dl and a normal LFT. Gastroduodenoscopy showed erythema of the stomach antrum, a 2cm X 2cm duodenal ulcero-proliferative growth at the junction of the first and second part of duodenum with suspicious thickening of the duodenal anterior superior wall and complete luminal and mucosal irregularity. Figure 1. Duodenal lesion biopsy revealed a high-grade intraepithelial neoplasm with associated duodenitis. His CEA 19.9 value was elevated to 63.4 U/ml. Histopathology examination was negative for malignancy.

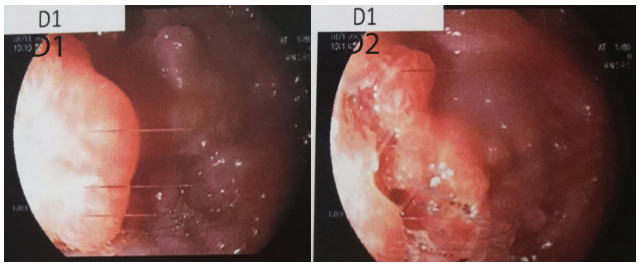

Figure 1. Duodenoscopy revealing growth at D1-D2 junction

A staging contrast-enhanced computerized tomogram (CECT) scan of the abdomen and pelvis showed a heterogeneously enhancing mass in the first part of the duodenum with suspicious loss of interface within the head of the pancreas and encasement of the gastroduodenal artery, suggestive of malignancy. Figure 2. Multiple lymph nodes in the hepatoduodenal ligament were found. Additionally, a dilated esophagus with gradual distal tapering suggestive of achalasia was affirmed. An auxiliary finding was a simple hepatic cyst.

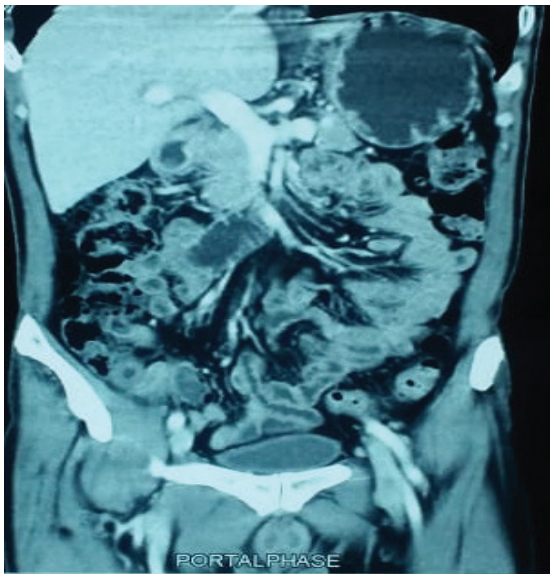

Figure 2. CECT abdomen: Suspicious thickening of Duodenum

Barium GI X-ray depicted a narrowing of the distal esophagus with upstream dilatation of lower and middle third esophagus with contrast hold up, these features were suggestive of achalasia cardia. Figure 3.

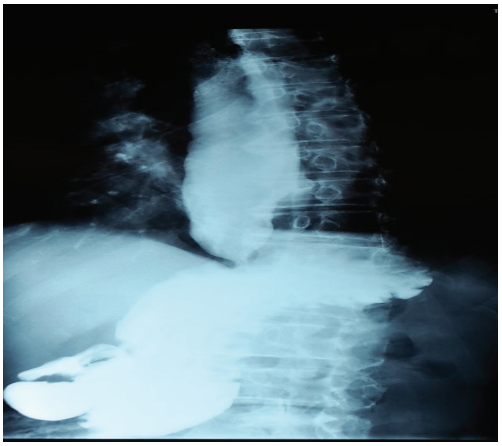

Figure 3 . Barium meal showing bird beak appearance of esophagus

His final diagnosis was primary duodenal carcinoma associated with an achalasia cardia. The patient underwent pancreaticoduodenectomy and Heller's cardio myotomy with subsequent Dor's fundoplication.

The total operation duration was approximately six hours, the patient's blood loss was around 300 ml. Peroperative findings were the constriction of the lower esophagus and the gastroesophageal junction with a proximal dilated esophagus, ulceroproliferative growth measuring around 3 cm x 3 cm with abutment of the gastroduodenal artery and the common hepatic artery was found in first part of duodenum. Figure 4. Additionally, there were multiple lymph nodes in the hepatoduodenal ligament within a low laying bifurcation of the proper hepatic artery. The main pancreatic duct was 1-2 mm, the pancreas appeared bulky and firm. The common hepatic duct had a diameter of approximately 3mm. Two drains were kept, one in the pancreaticojejunostomy site and one in the hepaticojejunostomy site.

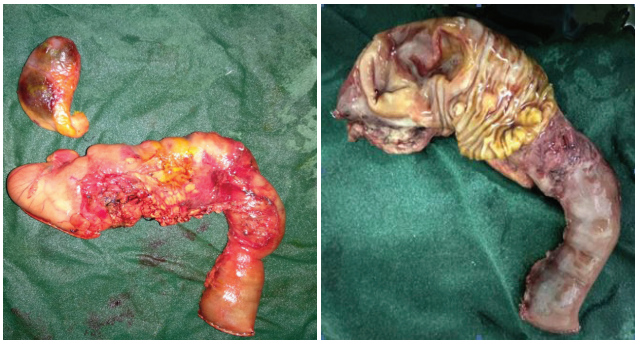

Figure 4. Resected specimen showing duodenal growth

He was kept nil per oral for 2 days with nasogastric decompression. Postoperatively, the patient received ceftriaxone for 5 days and octreotide for 3 days. Pain management was done with fentanyl infusion. Serum amylase and drain amylase from both drains were hinting at a pancreatic fistula grade A on the 3rd postoperative day. Apart from that, the postoperative history was uneventful. The patient was discharged on the 5th postoperative day.

Histopathological examination of the resected specimen revealed moderately differentiated adenocarcinoma at ampulla with a maximum dimension of 2.5cm. The tumor invaded the wall of the bile duct and duodenum up to muscularis propria. The entire resected margins were free of tumor. Perineural invasion was present while the lymphovascular invasion was absent. All 19 lymph nodes were free of tumor. The histopathological stage was pT3aN0.

On a month follow up, the patient showed improvement in his symptoms such as regurgitation, abdominal pain, and indigestion and was advised for adjuvant chemotherapy.

Discussion

Adenocarcinoma of the duodenum constitutes 73-90% of all primary malignant tumors of the organ, which accounts for approximately 45% of all small bowel adenocarcinomas.[5,6] The disease mostly affects the second part of the duodenum.[7] The association of achalasia cardia with duodenal malignancy is very rare. The patient presented with combined features of achalasia cardia and duodenal malignancy.

The duodenum is easily accessible by endoscopy, which implies the hypothesis that duodenal tumors should be diagnosed at an early stage. However, early symptoms are usually treated as ulcer disease, subjected to treatment without proper diagnostics, which often delays diagnosis and in most cases prevents radical management. The prerequisite is to maintain oncological vigilance and direct patients for an endoscopy every time symptoms persist for a long period of time. Upper gastrointestinal endoscopy with histopathological sampling is the diagnostic method of choice.

Similarly, a barium study can easily diagnose achalasia cardia. A staging CECT scan is required for assessing resectability. Added Heller's cardiomyotomy and Dor's fundoplication with pancreaticoduodenectomy can take care of both conditions.

Due to the rare location of the primary tumor, there is a lack of five-year survival results after radical procedures. Literature data results suggest a range between 20% and 90%. The number of reports on the efficacy of postoperative chemotherapy is negligible. Some studies showed the potential chemosensitivity of cancer cells to multiple drug schemes with 5 -fluorouracil leading to presumed prolonged survival.[8]

Conclusion

Duodenal cancer with achalasia cardia is a rare combination of rare diseases. Both diseases can be managed in a single sitting.

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