CASE REPORT

Papillary Adenocarcinoma in a Gastric Duplication Cyst

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Received: 16 April 2017 / Accepted: 30 October 2017 / Published online: 9 November 2017
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Abstract Gastric duplication cysts are rare and mostly present in the first year of life. In adulthood presentation is in the form of obstruction, ulceration, bleeding, fistulization etc. Malignancy is extremely rare with only 12 cases reported to date. We came across a gastric duplication cyst with papillary adenocarcinoma in a 63 year old man. He underwent cyst excision with radical subtotal gastrectomy. The awareness of such a condition made it possible for us to have a suspicion of malignancy preoperatively based on imaging and thus a radical surgery was performed. High index of suspicion is necessary to diagnose this condition preoperatively on CT scan. Literature review revealed that this is the first case to be reported from India.

Keywords Complication in gastric duplication cyst · Cancer in duplication cyst

Introduction

Gastric duplication cysts (GDC) are rare congenital anomalies, mostly presenting in the first year of life. Occasionally,

they are diagnosed in adults. Malignancy is extremely rare and only a few cases have been reported. We report the first case of malignancy in GDC in an Indian patient.

Clinical presentation

A 63-year-old male patient presented with complaints of vomiting since 15 days with one episode of hematemesis 3 days back. On physical examination, there was a palpable lump in the epigastrium. Upper gastrointestinal endoscopy was normal except for extrinsic compression at the antropyloric region. Scope was negotiable into duodenum with resistance. Ultrasonography (USG) revealed a large cystic lesion in the gastrohepatic ligament. He underwent computed tomography (CT) scan of the abdomen (Fig. 1). CT scan further revealed that the cyst showed no distinct planes with the inferior surface of the right lobe of liver. The stomach was hugely distended due to the pressure effect of the cyst on the antropyloric region causing partial gastric outlet obstruction. Internal soft tissue density polypoidal areas were seen within the cyst which showed intense enhancement in the post contrast study raising the suspicion of malignancy.

The differential diagnoses we considered were gastrointestinal stromal tumor (GIST) and GDC with malignancy. Intraoperatively, a large 10 × 8-cm-sized multiloculated cystic lesion was seen in the gastrohepatic ligament with adhesions to the antropyloric region and the inferior surface of the right lobe of liver. Frozen biopsy from the polypoidal component revealed adenocarcinoma. Cyst excision with radical subtotal gastrectomy, lymphadenectomy, and wedge resection of liver was performed. Postoperative histopathology was consistent with gastric duplication cyst with papillary adenocarcinoma (Fig. 2).

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Fig. 1 Coronal images of CECT abdomen showing distended stomach, with a lobulated, multicystic lesion (thin arrow) causing compression at the antropyloric region. The cystic lesion has thin enhancing walls and

few intraluminal papillary enhancing foci (broad arrow) which is highly suggestive of malignant transformation. The closest differential diagnosis is gastrointestinal stromal tumor

Discussion

The most common location for duplication cysts includes small intestine followed by esophagus and colon. GDC constitute approximately 4% of the cases [1]. The diagnostic criteria include (1) contiguous attachment to (but not necessarily luminal communication with) the stomach, (2) a smooth muscle coat that fuses with the muscle of the stomach, and (3) a mature or primitive gastrointestinal type epithelial lining. Two varieties are described, cystic (more common) and tubular. They are most frequently located at the greater curvature of the stomach followed by the posterior wall, lesser curvature, anterior wall,

and pylorus. Most of the GDC present in the first year of life, with vomiting and palpable abdominal mass. In majority of the cases, the diagnosis is incidental in adults. Other known complications include ulceration, bleeding, rupture, fistula formation, and pancreatitis from ectopic pancreatic tissue [2].

Malignancy in GDC is extremely rare with only 12 cases reported world wide [1–10, in English literature] [12, in Portuguese] [13, in Japanese] (Table 1). The median age of presentation is 56 years, but in two cases, malignancy is reported in young patients (25 and 28 years) reflecting the importance of timely detection of this entity [3, 8]. The average diameter is 8 cm [8]. The diagnostic and prognostic

Fig. 2 **a** Cyst wall composed of all the layers of duplication cyst (mucosa, submucosa, and muscularis) with denuded epithelium. H&E stain with $\times 4$ magnification. **b, c** Fragments of an infiltrating cellular neoplasm composed of papillary fronds with central fibrovascular core and lined by columnar cells with focal stratification. The cells exhibit moderate degree of nuclear atypia

(a) H & E stain with 4 X magnification

(b) 10 X magnification

(c) 20 x magnification

Table 1 Malignancy in GDC is extremely rare with only 12 cases reported world wide

SL no.	Author	Age	Sex	Location	Size	Surgery	Diagnosis	Histopathology
1	Barussaud et al. [1]	67	F	Adjacent to stomach with local carcinomatosis	NA	Excision of cyst and stomach	Postoperatively	Mixed Adenocarcinoma and squamous cell carcinoma
2	Kang HJ et al. [2]	56	M	Greater curvature of stomach	5.5 cm	Cyst excision with wedge removal of stomach	Postoperatively	Adenocarcinoma
3	Zheng J et al. [3]	25	M	Greater curvature of stomach	8 cm	Total gastrectomy with lymphadenectomy	Intra operative frozen section analysis	Adenocarcinoma
4	Horne G et al. [4]	40	M	Adjacent to stomach	12 cm	Total gastrectomy with splenectomy, distal pancreatectomy, wedge resection of left lobe of liver	Postoperatively	Neuro endocrine tumor
5	Moyo HW Jr. et al. [5]	64	F	Antrum of stomach	6 cm	Radical subtotal gastrectomy	Postoperatively	Well differentiated glandular carcinoma
6	Kuraoka K et al. [6]	40	male	Gastric fundus	7 cm	1: cyst excision, 2: proximal gastrectomy	Postoperatively	Adenocarcinoma
7	Coit DG et al. [7]	72	F	NA	4 cm	NA	NA	Mucinous papillary adenocarcinoma
8	Liu K et al. [8]	28	male	Gastric corpus	13 cm	Cyst excision	On follow-up: 7 months postoperatively [#]	Adenocarcinoma
9	Akio Yamasaki et al. [9]	42	F	Adjacent to greater curvature of stomach	10 cm	Complete resection of cyst with wedge resection of the stomach	Preoperatively on CT scan ^{##}	Moderately differentiated adenocarcinoma
10	Kondo J et al. [10]	75	male	Esophagogastric junction	6 cm	Proximal gastrectomy	Postoperatively	Adenocarcinoma
11	Present case	63	M	Lesser curvature of stomach near antropyloric region	10 cm	Cyst excision with radical subtotal gastrectomy with lymphadenectomy with wedge resection of liver	Intra operatively: frozen section of polypoidal lesions within the cyst	Adenocarcinoma

NA not available

[#] Biopsy at the time of surgery only revealed a gastric duplication cyst. However, there was spillage of the cyst contents into peritoneum during surgery. Seven months later, he presented with multiple intraabdominal malignant deposits with carcinomatosis peritonei^{##} Mural nodules within the cyst raised the suspicion of malignancy

implications of tumor markers still remain to be established. The most commonly reported tumor markers include CA19–9 and CEA. While in our case, both CA19–9 and CEA were within normal limits, Barussauda ML et al. [1] and Akio Yamasaki et al. [9] noted elevations in CA19–9, Liu K et al. [8] noted elevations in CA19–9, CEA, and CA-125 as well. Liu K et al. observed these elevations when patient presented with disseminated intra-abdominal malignancy. Adenocarcinoma is the most common histological type reported. The other rare varieties reported include mixed adenosquamous [1] and neuroendocrine carcinoma [5].

Preoperative diagnosis is difficult. In most of the cases reported in the past, the diagnosis of malignancy was postoperative. In one case, diagnosis was done retrospectively when patient came back with metastasis after 7 months of cyst excision [8]. As the duplication cyst may or may not communicate with the native stomach, it may be very difficult to have a biopsy with endoscopy alone. At present, CT scan remains the best imaging modality to diagnose GDC with malignancy. Differential diagnosis on computed tomography (CT) scan includes cystic lesions of nearby organs like pancreatic pseudocyst, mucinous cystadenoma of pancreas, GIST, and splenic cysts. In our case, GIST of the stomach was the closest differential diagnosis. Endoscopic ultrasonography (EUS) seems to be the best modality for demonstrating gastric duplications. EUS-guided fine needle aspiration (FNA) has been utilized in the past to distinguish GDC from GIST [11]. Few authors do recommend EUS to diagnose GDC with malignancy though none of them used it their respective cases (1, 2, 3).

The pathologic differential diagnosis includes gastrointestinal stromal tumor (GIST) and malignant transformation arising within a gastric teratoma. The clue to the diagnosis is the wall of a duplication cyst which is typically composed of a well-ordered arrangement of tissues that are native to the stomach, a fact which rules out a diagnosis of GIST (Fig. 2). The absence of a disordered arrangement of ectodermal, mesodermal, and endodermal tissues in the present tumor excludes the possibility of a teratoma.

Metastatic involvement was seen either at presentation or at follow-up imaging in 36.3% ($n = 4$). GDC with malignancy carries poor prognosis and the only effective treatment is aggressive surgical excision. Due to the rarity of this entity, no guidelines are available for the use of adjuvant therapy. In spite of adjuvant chemotherapy with 5 FU, cisplatin, and folinic acid administered by Barussauda ML et al., the patient died within 6 months with liver metastasis [1]. Our patient did not receive any chemotherapy.

Conclusion

Gastric duplication cysts with malignancy carry poor prognosis. Presentation is commonly in 5th decade but can occur even in the young. Preoperative diagnosis is extremely difficult. EUS may be useful in the work up of the patient. Radiologists should be aware of this condition as preoperative diagnosis will result in timely radical surgical management. We recommend that gastric duplication cysts should be excised whenever detected especially in adults even in asymptomatic cases.

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