Malignancy Arising in a Duplication Cyst of the Jejunum in an Elderly Man Presented as Intestinal Obstruction

A Case Report and Review of the Literature

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ABSTRACT

Enteric Duplication is a rare benign malformation that can occur anywhere along the digestive tract. It usually presents in infancy while adulthood presentation is rare. Malignant transformation is a very rare incident. In this paper; we report a case of malignant transformation in a jejunal duplication cyst in an elderly man who presented urgently with high small bowel obstruction. The case shall be presented, discussed along with literature review of this problem.

Keywords: Enteric duplications, Malignant changes, Small bowel obstruction, Endoluminal ultrasound.

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The term "Alimentary Tract Duplications" was first used by WE Ladd in 1940 to describe congenital malformations that involve the mesenteric side of the associated alimentary tract and share a common blood supply with the native bowel[1].

These malformations are rare, contain a normal gastrointestinal lining and can occur throughout the digestive tract but most commonly in the small bowel and most frequently in the lower ilieum around the ileocaecal region[2].

The etiology still remains unknown and most patients become symptomatic within the first year of life[3]. Reports in adults are extremely scarce in the literature and symptomatology is usually vague mimicking other more common gastrointestinal pathologies[4].

Carcinoma arising in an enteric duplication (ED) is a rare incident and only few cases have been reported in the literature[5].

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In here; we report an unusual case of a malignant ED cyst in a 77-year-old man presented urgently with high small bowel intestinal obstruction. The case shall be presented and the various aspects of diagnosis and management of this condition shall be discussed with appropriate literature review on this problem.

Case Report

A 78-year-old man was seen in the emergency department on the 11th of Feb. 2015 because of abdominal pain and repeated vomiting for two days duration. He gave a history of previous recurrent attacks of mild upper abdominal pain and his ultrasound (U/S) at that time reported a finding consistent with a mesenteric cyst (Figure 1: A and B). Otherwise; he has no history of any significant comorbidity. Surgery advised but the patient refused.

Clinically; the patient was moderately dehydrated with a remarkably tender distended silent abdomen and on digital rectal examination (DRE); the rectum is empty. The patient resuscitated and an erect abdominal film and necessary biochemical test were inconclusive.

On CT scan; there was evidence of a large intraperitoneal cystic lesion measuring around 14x12x7 cm with well-defined outlines consistent with omental or mesenteric cyst (Figure 2; A, B and C). There were multiple focal lesions in the liver for which a dynamic magnetic resonance imaging (MRI) was suggested for further evaluation.

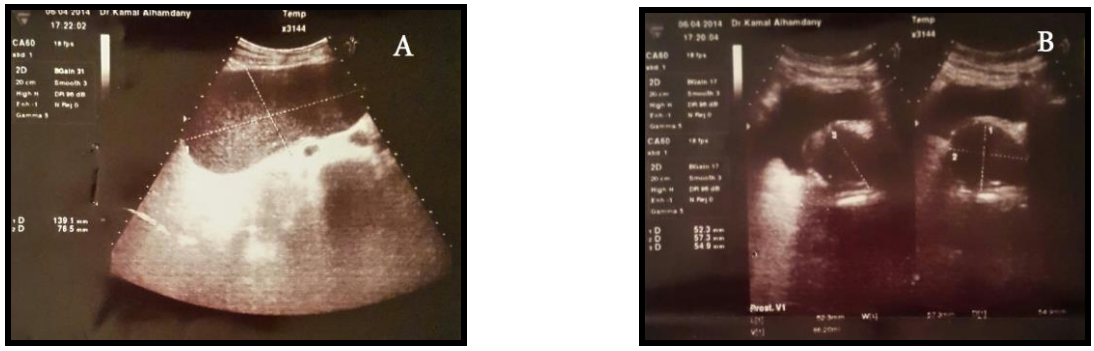

Figure 1 (A and B): U/S of the patient on Aug. 2014. An intraperitoneal well defined smooth outline iso-dense non-enhancing 13.2x13x7.3 cm mass containing unclear fluid and extending from the umbilicus down suggestive of a mesenteric cyst.

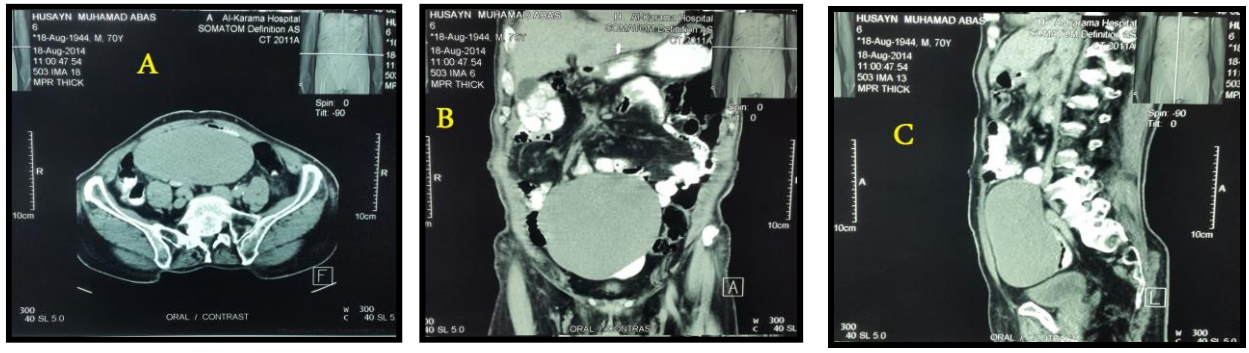

Figure 2 (A, B, and C): CT-scan with contrast the lumen of the adjacent small bowel is compressed and distal small bowel is collapsed.

The patient explored urgently and a relatively large rounded cystic mass could be identified in the mesentery of the jejunum around 50 cm from DJ junction. The wall is adherent to the adjacent small bowel and the outer surface is showing multiple small whitish nodules that can also be seen all over the peritoneum. The mass with around 30 cm of the adjacent small bowel was resected *en bloc* along with end-to-end anastomosis and a proximal feeding jejunostomy tube left in (Figure 3; A and B). After conclusion of the operation; the specimen was macroscopically inspected and the cyst was found filled with odorless thick brown material.

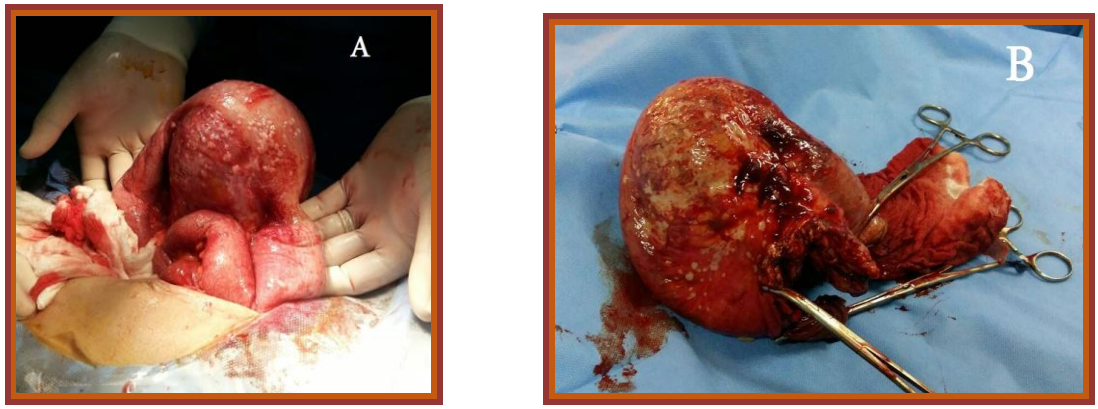

Figure 3 (A and B): The gross intraoperative appearance of the pathology and the resected specimen. The proximal jejunum is dilated due to obstruction secondary to extramural mass effect.

Histopathological assessment reported that the cyst wall is lined with flat layers of dysplastic epithelial cells with focal glandular differentiation. The wall is infiltrated by malignant pleomorphic epithelial cells forming strands, tubules and occasional glands invading the muscular layer of the wall associated with focal type of coagulative necrosis, (Figure 4). The picture is consistent with invasive poorly differentiated adenocarcinoma arising in a cystic duplication of the small bowel. The circumferential resected margin of cyst wall is invaded by the tumor. All the 13 regional mesenteric lymph nodes were involved by the tumor.

The patient had uneventful recovery and referred for adjuvant chemotherapy. Few months later, his condition deteriorated and he died of advanced intra-abdominal and pulmonary metastasis.

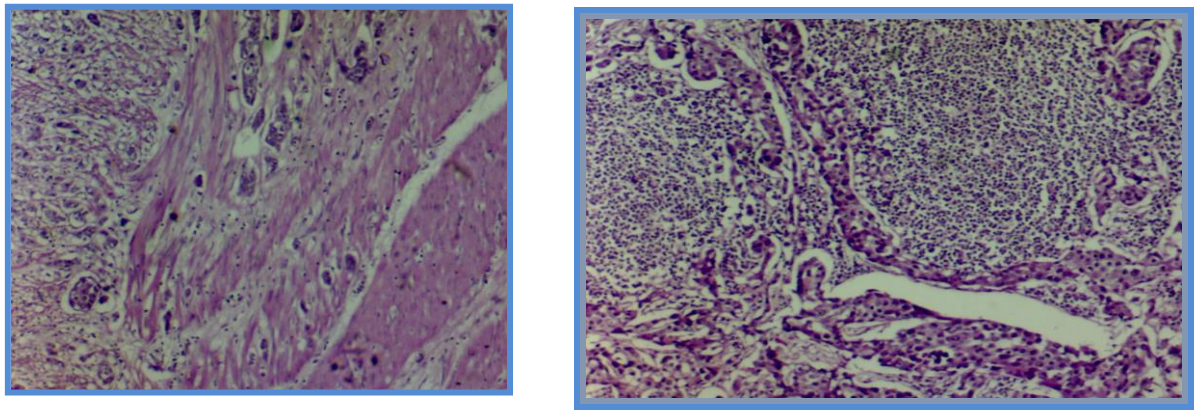

Figure 4: Poorly differentiated adenocarcinoma with evidence of lymphovascular invasion. The muscular layer of the cyst wall is invaded.

Discussion

EDs are malformations that usually present in infancy and although can occur anywhere along the digestive tract from the oral cavity to the rectum; the majority occurs intra-abdominally and over half of them are ileal duplications[6].

The etiology of EDs is unclear. Several theories have been postulated and the

earliest one stated that it is the result of abnormal recanalization after the solid epithelial stage of embryonic bowel development[7]. Another theory considered the etiology as persisting embryologic diverticula[8,9]. In 1971, an intrauterine vascular accident was proposed by Favara et al[10] as a possible pathogenesis and it had been considered as the most accepted theory. However; till now, no single theory can explain the pathogenesis of all known duplications[11].

Majority of patients with EDs become symptomatic within the first year of life; a palpable abdominal mass or symptoms of subacute intestinal obstruction are the most common. However; in older children, symptoms may mimic more common pathologies like volvulus, intussusception and Hirschsprung's disease[11,12].

In adult life; symptoms are more vague and may be confused with other more common pathologies at this period of life like appendicitis, intussusception, pelvic abscess, colonic or intestinal volvulus, diverticulitis and bowel cancer, however; in adults, EDs are most commonly diagnosed when complications like bowel obstruction, perforation or bleeding occur[13,14].

The majority of EDs are isolated and cystic in structure. Reports of tubular EDs are less common, however; both could be associated with other malformations like intestinal malrotation[15], genitourinary[16] or spinal malformations[17].

Prior to surgery; it is difficult to diagnose an ED because of non-specificity of symptoms and presentation, however; ultrasound (U/S), CT scan and magnetic resonance imaging (MRI) have all been useful[18].

U/S can depict the characteristic location adjacent to the bowel and the two-layer wall of an ED. CT scan can be useful for delineation and assessment of the relation to the bowel and especially in tubular EDs, CT scan with contrast may give a clearer definition. On MRI; EDs present with heterogeneous signal intensity on T_1- and homogenous signal intensity on T_2- weighted images. In tubular duplications, barium examination may be diagnostic if not contraindicated. There is also a role for technetium scanning in diagnosis[19, 20].

Recently; there has been a focus on the role of endoscopic U/S (EUS) for the evaluation and diagnosis of ED. It is said to be the diagnostic tool of choice as it can distinguish between solid and cystic component of the lesion and it can establish cyst location relative to surrounding tissue[21]. On EUS; the cyst showed as anechoic homogeneous lesions with regular margins arising from the submucosal layer or extrinsic to the gut wall. The wall is usually consist of 3-4 layers and the internal contents may be anechoic or hypoechoic. It may show septation, fluid level, debris and detached ciliary tufts which could be diagnostic[22]. In addition; ED cyst may demonstrate peristalsis that appears as ring contractions with concentric contractions of the cyst wall. Peristalsis in a juxta-enteric cyst is specific for ED and can be a diagnostic feature[23].

There has been a lot of controversy concerning performing fine needle aspiration cytology (FNAC) under EUS guidance for lesions suspected to be an ED cyst[24]. EUS-guided FNAC may facilitate definitive diagnosis, may rule out more ominous lesions and may be therapeutic if the cyst is evacuated and observed for recurrence. The controversy is based on the fact that these lesions can become infected with significant consequences[25,26].

Carcinoma arising in ED is an extremely rare incident and only few cases have been reported in the literature including different pathologies like carcinoid tumor[27], squamous cell carcinoma[28,29] and the more common adenocarcinoma[30]. Malignant changes in EDs are described most frequently in small bowel[30-32] followed by colonic and rectal duplications[33-36]. There have been also reports about carcinomas arising in duplications of the duodenum and the

stomach[37-39]. Metastatic adenocarcinoma from an ED had also been reported[40]. Heterotopic mucosa of gastric or pancreatic origin is a relatively common finding in histological examination of EDs and malignant changes had also been reported[41].

Once the diagnosis of an ED cyst is established, treatment can vary depending on the presence of symptoms. In symptomatic patients, surgical resection is highly indicated for relief. In asymptomatic patients; surgical resection is controversial and observation, EUS-guided FNAC plus observation or immediate resection, had all been advocated. Since there have been reports of stable ED Cysts on EUS surveillance, this may be a suitable method of outpatient follow up and surgical resection can be considered if patient develops symptoms. In any case; surgery versus no surgery for asymptomatic EDs is likely to remain controversial until more understanding about the time course and risk factors associated with malignant transformation, is available[42-44]. It should be remembered that due to the rare presentation and unspecific symptoms of EDs, malignancy is commonly diagnosed at an advanced stage and probably with a high rate of lymph node metastatic disease. The mode of metastasis is similar to that of primary small bowel cancer and a curative resection may be difficult to be attended, thus, the prognosis is generally poor once malignant changes have occurred[42-44].

Conclusions and recommendations: The presented case stresses the importance of high index of suspicion towards innocent appearing intra-abdominal cystic lesions in elderly people. Surgery, once appropriate, is highly indicated early in the course of such lesions. Delayed cases carry a poor prognosis. Documenting unusual presentations of rare cases is highly recommended.

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