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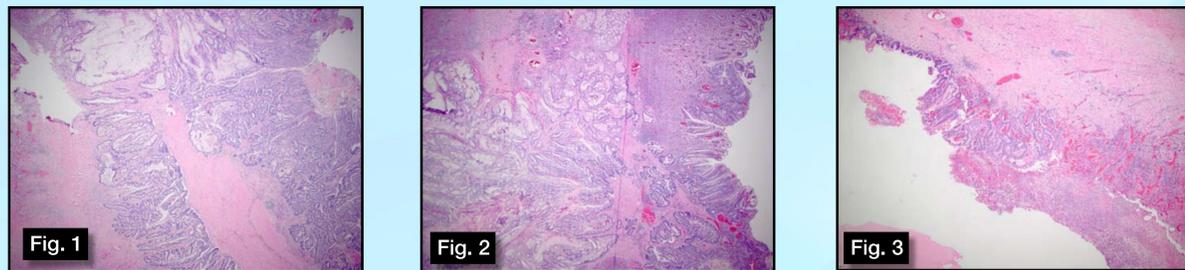
A Case Report of Enteric Duplication Cyst: A Unique Presentation With Multiple Rare Associations

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Case Presentation:

A 59-year-old African American male presented with icteric sclerae, dark urine, dyspnea, abdominal pain and fatigue for 1 month. His medical history was significant for type 2 DM and untreated hepatitis C. Routine blood work showed Hb 6.8 g/ dL with baseline at 13.5 g/ dL about 5 months ago. WBC count and platelet count were normal. Other tests showed MCV 112 fL, reticulocyte count 20.6 %, LDH 1423 IU/L, total bilirubin 10.1mg/dL and undetectable haptoglobin level. The direct antiglobulin test (DAT) was positive for immunoglobulin G and complement C3d. He had negative HIV and autoimmune panel. CT scan abdomen showed spherical mass in the right abdomen measuring 6.6 cm x 6.6 cm x 8.3cm abutting the inferior aspect of the duodenum. This was followed by endoscopic ultrasound revealing a complex necrotic-appearing, well-circumscribed structure 7 x 6 cm adjacent to the duodenum which did not emanate off of the pancreas. The cytology ruled out malignancy. He was further started on prednisone for W-AIHA. His hemoglobin improved with steroids however his hemolysis flared with positive DAT every time on steroid weaning. His active untreated Hepatitis C with heavy viral load precluded treatment with rituximab or immunosuppressants. He was a poor splenectomy candidate due to his underlying chronic liver disease. He was continued on low dose steroids for 18 months with stable hemoglobin. He developed osteoporosis and avascular necrosis of hip secondary to long term steroids. Follow up CT scan showed stability of the abdominal lesion.

At 2 years, he presented with complaints of recurrent abdominal pain and 15 pound weight loss. CT abdomen showed increase in size of the complex retroperitoneal mass with thickened wall and soft tissue stranding. He underwent upper endoscopy and biopsy revealing intra-mucosal adenocarcinoma of the duodenum. He further underwent whipple resection with final pathology showing stage IIA (pT3 N0 M0) duodenal adenocarcinoma with negative margins. The tumor involving the duodenum showed continuity with the underlying 6.5 cm cyst. The cyst was predominantly necrotic however there were foci showing intact small bowel mucosa consistent with enteric duplication cyst. At 2 month post-surgery he was successfully weaned of steroids and had complete resolution of hemolytic anemia with negative DAT.



Figures 1, 2, 3: Invasive moderately differentiated adenocarcinoma, 2.5 cm, involving duodenum, with invasion through muscularis propria and into underlying necrotic duplication cyst.



Figure 4: Complex right retroperitoneal mass measuring 7.3 x 7 x 8.4 cm, slightly increased in size from 6.6 x 6.6 x 8.3 cm two years ago. Also shows new wall thickening with mild soft tissue stranding in the adjacent fat.

Figures 5, 6, 7: The tumor involving the duodenum shows continuity with the underlying 6.5 cm cyst. The cyst shows predominantly necrotic contents; one focus, however, reveals intact small bowel mucosa consistent with an enteric duplication cyst.

Discussion:

Gastrointestinal duplication cysts are rare congenital anomalies of the alimentary canal that usually present in childhood with an incidence of 1 in 100,000 cases per year.¹ Duodenal duplication cyst forms about 2-12 % of all the gastrointestinal duplication cyst.¹ GI duplication cyst is defined by 3 Rowling's criteria: (1) the wall of the duplication is in continuity with one of the duplicated organ; (2) the cyst is surrounded by a smooth muscular layer; and (3) a layer of digestive mucosa is present, more often typical or heterotopic as gastric mucosa, colonic mucosa, bronchial or pancreatic structure.^{2,3} Duodenal duplication cyst consists of submucosa, muscularis propria, and a duodenal epithelial lining with close attachment to the alimentary tract.

The case presents two very rare associations of enteric duplication cysts: W- AIHA and secondary malignancy. Hemolytic anemia is commonly associated with lymphoproliferative neoplasms, drugs, connective tissue diseases, infections and are very rarely reported in association with ovarian dermoid cyst and mesenteric dermoid cysts.⁵ In our patient, AIHA was initially presumed to be hepatitis C induced, however the temporal association of hemolytic anemia resolution with surgery favors a strong possibility of association between the two. Though carcinomas are known cause of secondary autoimmune hemolytic anemia, since our patient presented with progressive disease 2 years later, this would preclude definite conclusion on malignancy as the underlying etiology for W-AIHA. Recognition of this association can be invaluable as it presents an opportunity for definitive management and spares the need for potentially risky immunosuppressive therapy and splenectomy. Treatment of the primary disease results in remission of AIHA with DAT reverting to negative. Reemergence of DAT positivity in these patients warrant thorough investigation to rule out relapse.

Carcinomas arising in duplication cysts are extremely rare complications and only few cases (<30 cases) have been reported in literature including carcinoid tumors, squamous cell carcinomas, and adenocarcinomas.⁴ Malignant transformation is more commonly reported in small bowel, colonic and rectal duplication cyst with only two cases reported from duodenal duplication cyst.⁴ Due to the rarity and nonspecific presentation, early identification of malignant transformation could be most often missed. Due to early lymphatic spread and less chance of curative en-bloc resection, these tumors are considered to have poor prognosis.⁴ Fortunately our patient had no lymph node metastasis and he is doing well without adjuvant therapy. Duplication cysts identified in pediatric population is mostly benign, however malignant transformation is mainly identified in the older population. Therefore identification of duplication cyst in an older individual should trigger appropriate surveillance in these patient population.

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