Primary Appendiceal Gastrointestinal Stromal Tumor: A Case of an Incidental Tumor

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Introduction/Objective: Gastrointestinal stromal tumors (GISTs) are rare with an approximate prevalence of 2%. Although rare, they are the most common mesenchymal neoplasm of the gastrointestinal tract. They commonly occur in the stomach (50-60%), small intestine (20-30%), large intestine and esophagus (<10%). GISTs arising in the appendix are very rare, with less than 20 cases reported in the English literature to date. The clinical presentation varies from appendicitis-like symptoms to incidental findings on imaging, during surgery for other diseases or at autopsy. The differential diagnosis of GIST from other stromal tumors is often difficult on hematoxylin and eosin (H&E) examination alone. The use of immunohistochemistry (IHC) plays a key role in confirming the diagnosis. GISTs can be graded based on a four-tier risk stratification system, including high, intermediate, low and very low risk, taking size tumor and mitotic activity into consideration. Most of the previously reported cases of primary appendiceal GIST were low or very low risk but one case of malignant GIST arising in the appendix has been reported.

Methods: We present the case of a 65-year-old male with a slightly thickened appendix and fluid in the distal aspect incidentally found on computer tomography (CT) scan for diverticulitis workup. An appendectomy with adequate was performed. The gross specimen revealed a 0.2-0.4 cm thick wall with no gross perforations or masses. Histologically, the tumor measured 0.8 cm and was composed of spindle cells with no nuclear atypia and absent to low mitotic activity. The IHC profile of the tumor was positive for CD117, DOG1 and CD34. A diagnosis of Gastrointestinal stromal tumor, low grade was rendered.

Conclusion: We endeavor to highlight the importance of considering GIST in the differential diagnosis in patients with nonspecific symptoms and/or atypical image findings of the appendix especially in ruling out a possible malignant GIST.

A Case of Small-Cell Neuroendocrine Carcinoma of the Gallbladder in a Background of High-Grade Biliary Intraepithelial Neoplasia

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Introduction/Objective: Neuroendocrine carcinoma (NEC) of the gallbladder is a very rare neoplasia comprising 4% of all malignant gallbladder neoplasms. Most of NECs show molecular accumulation of TP53, while KRAS mutations are rare. Approximately 40% of Biliary intraepithelial neoplasia (BilIN) cases are associated by KRAS mutations as an early molecular event, whereas TP53 mutation appears to be a late molecular event.

Methods: We report a case of a 62-year-old male who presented to emergency department for evaluation of abdominal pain associated with vomiting. Imaging studies demonstrated a distended gallbladder with small amount of pericholecystic fluid along with 5 mm calculus at the gallbladder neck consistent with symptomatic cholecystitis and acute cholecystitis. No mass lesion was radiologically identified. A laparoscopic cholecystectomy was performed.

Grossly, there was a thickened area of mucosa at the distal body-fundus, measuring 5.0 x 4.5 cm. Multiple calculi were present. On histology, thickened area showed sheets and nests of moulded small cells with hyperchromatic nuclei, brisk mitotic activity and confluent necrosis. A diagnosis of small-cell NEC is established, and confirmed by positive immunoreactivity to neuroendocrine markers (CD56, Synaptophysin) and epithelial markers (CK7 and CAM5.2). Multiple foci of high-grade BilIN were noted. Subsequent entire submission of the gallbladder revealed no adenocarcinoma present. The pathologic stage was pT2a pNX.

Conclusion: In conclusion, this rare gallbladder carcinoma is found incidentally in a patient with acute cholecystitis symptoms and no clinical proof of mass lesion. In addition, there is a background of high-grade BilIN, which shares the molecular pathway of small-cell NEC. Therefore, awareness of such coexistence of these two pathologic entities in the same gallbladder, is essential to alert pathologists to look for the poorly-prognosis small cell NEC, whenever BilIN is encountered in random section of a gallbladder.

A Rare Presentation of Pseudomembranous Colitis in a COVID-19 patient

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Introduction/Objective: Recent reports suggest that though COVID-19 is predominantly a respiratory pathogen, one of its presenting features can be gastrointestinal symptoms.
We hereby present a case of a female with COVID-19 infection whose hospital course was complicated by colonic pseudo-obstruction caused by pseudomembranous colitis resulting in an emergent hemicolectomy.

**Methods:** A 59-Year-old female with history of hypertension, diabetes, and breast cancer post mastectomy presented with pneumonia and was confirmed to be COVID-19 positive. She was admitted to the hospital and was treated with Azithromycin for 6 days. Patient developed constipation on day six of hospitalization and started having abdominal pain on day eight with elevated WBC count. Imaging showed distension of cecum and proximal colon. She underwent exploratory laparotomy which revealed a necrotic appearing cecum that was massively dilated and had a serosal tear. These findings prompted emergent hemicolectomy with loop ileostomy. Grossly cecum was black/green, dilated, thin walled with a 5 x 5 cm yellow green raised plaques. Microscopy of the plaques revealed focal erosion of colonic mucosa with overlying acute inflammatory cells, fibrin deposits, mucus, and necrotic epithelial cells consistent with pseudomembranous colitis.

**Conclusion:** Review of literature shows no reported cases of intestinal pseudo-obstruction due to pseudomembranous colitis in a COVID-19 patient. Not only this, but there are also only a limited number of case studies of pseudomembranous colitis presenting as intestinal pseudo-obstruction without diarrhea. Though this patient’s presentation could be from Clostridium difficile infection secondary to Azithromycin, it is not a common antibiotic to cause this. Also, one of the known causes of pseudomembranous colitis is ischemia. Given that COVID-19 infection is a pro-thrombotic condition, possible ischemia secondary to COVID-19 infection related coagulopathy should also be a consideration.

**Hirschsprung’s Disease: Two Cases of Total Intestinal Aganglionosis**

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**Introduction/Objective:** Hirschsprung’s disease is a disorder associated with an absence of ganglion cells in a segment of the bowel. Total colonic and small bowel aganglionosis occurs in less than 1% of all Hirschsprung’s disease patients. Even rarer is the finding of Haddad syndrome, the coexistence of congenital central hypoventilation syndrome and Hirschsprung’s disease. Congenital central hypoventilation syndrome has approximately 1,000 reported cases, with colonic aganglionosis being associated with 15-20% of those cases. The prevalence of Hirschsprung’s disease with congenital renal anomalies may be another underreported finding.

**Methods:** Here, we report two rare cases of Hirschsprung’s disease with significant extension in the small bowel. One case involves a 38-week gestational age male infant admitted for apnea and another case involves 39-week gestational age female infant admitted for vomiting.

**Results:** Both infants had rectal suction biopsies confirming the absence of ganglion cells. The male infant had mapping biopsies which histologically showed hypertrophic nerve fibers and an absence of ganglion cells in the colon extending up to 70 cm proximal to the ileocecal valve. Calretinin immunostaining confirmed aganglionsis. Genetic testing showed a PHOX2B gene mutation carrying 32 polyalanine repeat mutations, confirming a diagnosis of congenital central hypoventilation syndrome coexisting with Hirschsprung’s disease. The female infant had intraoperative frozen sections that identified ganglion cells throughout the colon. On permanent sections, there was an absence of ganglion cells in the upper rectum extending up to 65 cm proximal to the ileocecal valve. She also was noted to have agenesis of the left kidney and a urachal remnant. Genetic testing was negative for RET mutations.

**Conclusion:** Overall, these cases provide further information on a rare variant of Hirschsprung’s disease that includes significant portions of the small bowel. Additionally, this study adds to the documented reports of Haddad syndrome and the connections between renal anomalies with Hirschsprung’s disease. Last, this series alludes to the difficulties of frozen section diagnosis of this disease.

**A Case Of Pancreatic Hamartoma Mimicking Malignancy: An Uncommon Pitfall**

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**Casestudy:** Hamartomas are benign, disordered, tumor-like growth of cellular constituents resembling the tissue of its origin. Hamartomas are usually seen in lung, heart, kidney and spleen. Pancreatic hamartomas (PH) are extremely rare, accounting for <1% of all hamartomas. PH occurs at any age (median: 50 years) without gender predilection. PH presents as single or multiple, solid and/ or cystic mass composed of exocrine tissues. Admixed neuroendocrine cells may be seen, but well-formed islets are unusual. PH stroma is typically positive for CD34 by immunohistochemistry. We present a case of a PH resected for the clinical suspicion of malignancy, with the final diagnosis established postoperatively.

The case is that of a 74-year-old male with an incidental 2.3 x 1.7 x 1.1 cm hyperenhancing solid mass of the pancreatic