A0649- Goblet Cell Adenocarcinoma Presenting as Appendicitis

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Introduction

- Goblet Cell Adenocarcinoma (GCA) of the appendix is a very rare subtype of appendiceal tumor with an estimated incidence of 1 per 2 million people.
- It is not only rare but is also unique in origin. Most cancers arise from a single cell line but GCA of the appendix is a hybrid of neuroendocrine cells and epithelial cells.

- . The aim of this abstract is to provide an overview of this unique clinical entity.

Case Description

- A 62- year old female with past medical history of hypertension, diabetes mellitus and hyperlipidemia presented to the ED with a one day history of right lower quadrant abdominal pain that had migrated from the periumbilical area. The pain was associated with non bloody vomiting.
- She was stable on arrival and her physical exam demonstrated right lower quadrant tenderness along with positive Rovsing and Obturator signs.
- Labs were remarkable for leukocytosis with left shift. CT scan of the abdomen revealed an inflamed appendix and the patient was taken for laparoscopic appendectomy.
- Pathology of the surgical specimen revealed a well differentiated T1NxMx GCA with negative margins and no lymphovascular or perineural invasion. A decision was made to proceed with a right hemicolectomy for staging and treatment.
- Pathology sent after right hemicolectomy revealed no residual GCA with 21 lymph nodes negative for metastasis. Surgical margins were negative as well.

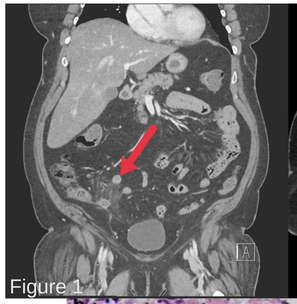
Figure 1

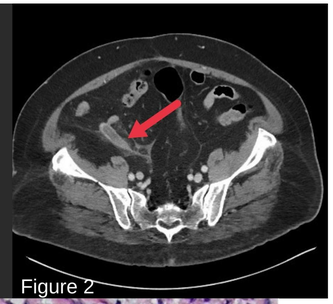
Figure 2

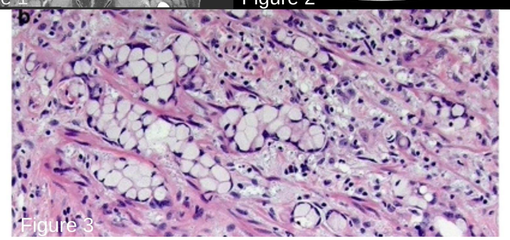
Figure 3

Figure 1 and 2- CT scan of the abdomen showing distended appendix with appendiceal wall thickening and hyperemia, as well as trace periappendiceal edema. Figure 3- Histologically conventional Goblet cell adenocarcinoma has a “crypt cell pattern” characterized by small round clusters of goblet cells in acinar configuration, resembling colonic crypts. [Pathology slide from Mod Pathol. 2016;29(10):1243-1253. doi:10.1038/modpathol.2016.105)]

Discussion

- First described in 1969, GCAs mostly occur in the 5th and 6th decade of life with equal prevalence in both genders.
- GCAs most commonly present as acute appendicitis but can also present with abdominal pain associated with an abdominal mass and weight loss. They can metastasize to the peritoneum, omentum, abdominal wall, and ovaries.
- Most GCAs are diagnosed incidentally on pathological examination of the inflamed appendix after an appendectomy.
- The mainstay of treatment is surgical resection with the addition of adjuvant chemotherapy for node-positive (stage III) disease or higher.
- Given the fact that this is a rare condition, there is a lack of consensus regarding the extent of surgical resection. For stage I disease, appendectomy alone is sufficient, but higher stages may require hemicolectomy.
- Prognosis depends on the grade and stage at diagnosis, with one retrospective analysis reporting 5-year survival rates of 100%, 76%, 22%, and 14% for stages I, II, III, and IV, respectively.

References

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