

Answers to Medical Quiz

A Lady with Mucous Discharge Per Rectum

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A1. Structure involved is the caecum. There is a thin walled, unilocular, pseudocystic mass at the base of the caecum containing thick jelly-like mucus. Also the distal part of the appendix is abnormally dilated, though its mucinous content has been removed to illustrate the smooth inner surface.

A2. The diagnosis is benign appendiceal mucinous cystadenoma, other clinical differential diagnoses might include:

- Caecal mucinous carcinoma.
- Mucocoele.
- Abscess formation secondary to a cecal diverticulum.
- Phlegmon mass secondary to a localized ruptured appendicitis. **
- Metastatic primary ovarian mucinous tumour.
- Caecal Tuberculosis.

A3. See the discussion below.

DISCUSSION

The primary benign mucinous cystadenoma of the appendix has an unusual secondary dissection of the mucinous content into the caecal wall forming a pseudocystic mass close to the appendiceal base.

Microscopically, the caecal cyst is formed by a thin fibrous wall, has no lining epithelium and its outer surface, which face the caecal lumen is covered partly by ulcerated colonic mucosa. The dilated appendiceal part is lined by atypical low grade mucus-secreting epithelium that shows focal papillary configuration. The lumen at the proximal appendiceal part was occluded by transmural fibrosis. In the face of the proximal appendiceal lumen obstruction and as a result of increased intraluminal pressure distally, the mucous penetrates and dissect into the wall similar to diverticulosis. In this case it has reached the caecal wall, and as a result it produced a pseudocyst just underneath the caecal mucosa. In other circumstances, similar mucous dissections might present as a periappendiceal or retroperitoneal mass with or without secondary perforation and subsequent development of pseudomyxoma peritonei, see below.

The term Mucocele is not an entity but rather a generic non-specific expression, often used by surgeons to denote accumulation of mucinous material in any given organ or structure. It tells nothing about the underlying pathologic process leading for its formation¹⁻².

In the appendix, there are several main pathologies that can give rise to similar clinical presentations; first simple luminal occlusion due to Inflammatory or benign conditions such as endometriosis, cystic fibrosis or obstructing carcinoid tumour; in these conditions the epithelium is usually flat and atrophic. Second is localized mucosal hyperplasia, much similar to hyperplastic colorectal polyps, and third entity true mucinous neoplasms that have specific morphologic, histochemical and ultrastructural features. The last entity includes adenomatous polyps, benign mucinous cystadenomas, and malignant mucinous cystadenocarcinomas¹.

Points of clinical importance are the co-association of such appendiceal mucinous neoplasms whether benign or malignant with other synchronous or metachronous neoplasms of similar nature in the ovaries or colon¹.

The gross appearance of benign, borderline and malignant mucinous neoplasms can be similar, however, microscopically, identification of invasion of the appendiceal wall or any nearby structure by atypical malignant epithelial cells or their presence in the peritoneal mucinous deposits even they appear bland, should give away the clear cut distinction^{1,2}.

Similarly, the site of origin might be difficult to ascertain in advanced malignant conditions that usually tend to involve the right side of the abdomen. In such situations, immunohistochemical markers can be of great help in sorting out this problem especially the utilization of cytokeratins CK7 and CK20 that stain ovarian and large bowel mucinous tumours respectively³. Galectin 4 and Meprin alpha are two recently described additional markers, which had been claimed useful in differentiating primary from secondary mucinous adenocarcinomas of the ovary³.

In this case the patient has underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy 15 years ago!

Appendectomy is the treatment of choice if the mucocele is located on the tip of appendix, however if its on the base or encroaching to the cecum (like in our case) a right hemicolectomy is warranted to rule out the possibilities of mucous cystadenocarcinoma for a curative surgery especially without the benefit of a frozen section.

REFERENCES

1. Juan Rosai. Rosai and Ackerman's Surgical Pathology textbook, volume 1, Ninth edition: London, Mosby: 2004; 761-5.
 2. Harry S. Cooper. Intestinal Neoplasms. In Sternberg's diagnostic surgical pathology textbook, volume 2, fourth edition: Stacey E. Mills (senior editor). Philadelphia: Lippincott Williams and Wilkins:2004; 1576-7.
 3. Heinzelmann-Schwarz VA, Scolyer RA, Scurry JP, et al. Low meprin alpha expression differentiates primary ovarian mucinous carcinoma from gastrointestinal cancers that commonly metastasise to the ovaries. J Clin Pathol 2006, 5; [Epub ahead of print]
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