

Answers to Medical Quiz

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A.1. MRI of abdomen and pelvis

The findings were:

1. A cystic lesion, about 10 cm in length
2. The lesion is at the level of the right colon
3. The lesion was extending from the right iliac fossa to the lower pole of right kidney

A.2. The differential diagnosis includes:

1. Appendicular swelling which could be appendicular mass or abscess, mucocele of the appendix, appendicular adenoma or adenocarcinoma (mucinous or colonic type), carcinoid tumor (classic type- tubular or goblet cell carcinoid) and lymphoma of the appendix
2. Duplication cyst of the right colon (congenital)
3. Right hydroureter
4. Right hydrosalpinx or ovarian cyst
5. Mesenteric cyst

A.3. There are no specific laboratory tests, which could finalize the diagnosis except in carcinoid tumor and adenocarcinoma of the appendix. The tumor markers are the following:

- 5 hydroxytryptamine in serum and 5 hydroxyindole acetic acid in urine
- Carcinoembryonic antigen (CEA)

A.4. The diagnosis was mucocele of the appendix confirmed by histopathology

DISCUSSION

Mucocele of the appendix is a descriptive term, which implies cystic dilatation of the appendix filled with abnormally accumulated mucus. It is uncommon and the incidence of mucocele in appendectomy specimens is 0.2 to 0.3%¹.

The mean age of presentation is 55 years with female preponderance of 4:1. In women with appendiceal mucocele, the ovaries must be thoroughly examined for cystic tumors².

About 25% of mucocèles are asymptomatic and discovered incidentally as the case in our patient. Other clinical features include right lower quadrant pain, a palpable abdominal mass, sepsis, urinary symptoms and rarely intermittent colicky pain provoked by intussusception of the mucocele into the cecum².

Preoperative diagnosis of mucocele is important to reduce the possibility of rupture and development of pseudo myxoma peritonei and to predict malignant transformation. Plain X-ray of mucocele may appear as a soft tissue mass in the right lower quadrant with or without calcification. Sonography is valuable in demonstrating mucocele; it might show a large hypoechoic mass. CT and MRI are the

avored imaging techniques for diagnosing appendicular mucocele³. Mucoceles up to 40 cm x 24 cm x 20 cm have been reported³.

Mucoceles were first described by Rokitansky⁴. The mucocele was thought to be the result of obstruction with distal accumulation of mucus. Obstruction was commonly post inflammatory but could also be due to fecaliths, congenital obstruction of Gerlach's valve or extramural compression. Evidence from pathological examinations suggests that mucosal atypia or neoplasia is the cause of mucocele formation⁵.

Higa et al classified these lesions into three groups, focal or diffuse mucosal hyperplasia without atypia, which was found in our patient, mucinous cystadenoma and mucinous cystadenocarcinoma⁶.

The commonest type is mucinous cystadenoma (about 60%), which exhibits focal or diffuse conversion of the mucosa into neoplastic epithelium. Most patients are asymptomatic and have an excellent prognosis.

Mucinous cystadenocarcinoma though less common (10-15%) but are often symptomatic⁶. Complications of mucocele include pseudomyxoma peritonei and intussusception. Pseudomyxoma peritonei is characterized by implants of mucinous epithelium on the peritoneal surfaces and mucus accumulation within the peritoneal cavity. It usually follows rupture of a benign or malignant mucocele. Adhesions and intestinal obstruction are the most frequent complications of this condition. If pseudomyxoma peritonei is found in the setting of mucinous cystadenocarcinoma, the prognosis is poor with 20% five-year survival rate⁷.

Although mucocele is a benign lesion, it can undergo a malignant transformation; therefore, the treatment of choice is appendectomy and careful inspection of the abdomen for any associated colonic neoplasms or ovarian tumors. If malignancy is suspected, the procedure should be converted to hemicolectomy. It is important to be very careful when mobilizing the tumor to avoid opening the mucocele and causing dissemination. If the mucocele perforates in the peritoneal cavity, it may lead to the development of a serious complication called pseudo myxoma peritonei⁸.

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