MUCINOUS NEOPLASM OF THE APPENDIX: A CASE REPORT

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ABSTRACT

A 28-year-old Indian male reported abdominal pain and vomiting for two days, but no fever. Upon examination, a doubtful non-tender mass was palpable in the right lower abdomen. No other morbidity was noted. The hematological tests were normal. The patient was advised an ultrasonography of the abdomen, which revealed an intra-abdominal hypoechoic/sonolucent mass on the right side and bilateral renal calculi. A computed tomography (CT) scan of the abdomen and pelvis was performed and it showed the enhancement of a low-attenuation, faintly calcified tubular mass in the right iliac fossa. The case reported at our center is most likely to be that of mucinous cystadenoma (tumor of the appendix) rather than mucocoele. The patient subsequently went to India, where colonoscopy and the Carcinogenic Antigen Test (CEA) were performed. The results were normal and he was operated. The cystic mass was resected with appendectomy. Histopathologically, it was diagnosed as mucinous cystadenoma of the appendix. A six-month follow-up did not reveal any recurrence. Mucinous cystadenoma presenting as an appendiceal mucocoele neoplasm is rare. Most of the times, it is incidentally detected during the course of other abdominal surgeries.

Keywords: Mucinous cyst adenoma, appendix, appendiceal mucocoele


INTRODUCTION

Radiological investigations play a major role in the diagnosis and management of neoplasm of the appendix. Mucinous neoplasms of the appendix range from simple mucocoele to complex pseudomyxoma peritonei. Primary neoplasms of the appendix are diagnosed during the course of other abdominal surgeries or when a patient is operated for appendicitis and the specimen is sent for histopathology.

In 1842, Rokitansky first described, clinically and radiologically, the mucocoele of the appendix. The term ‘mucocoele’ refers to the cystic dilatation of the appendix filled with mucin, resulting from the proximal obstruction of the appendix lumen. It usually presents as appendicitis in a middle-aged person. If there is a delay in diagnosis, the mucinous cystadenoma may rupture and result in pseudomyxoma peritonei. Neoplasms of the appendix account for only 0.4% of gastrointestinal tumors. Based on the CT scan, we presented a case of mucinous cystadenoma of the appendix. Surgery and histopathology confirmed this.

CASE REPORT

A 28-year-old man, admitted to Thumbay Hospital, Fujairah, reported that he had abdominal pain and vomiting for two days. No fever or other comorbidities were reported. According to the clinical examination, his BP was 130/80 mmHg and the pulse rate was 70 per minute. He had a normal respiratory rate. Nothing abnormal was detected during the examination of the central nervous system (CNS), central venous system (CVS) and chest. While examining the abdomen, a doubtful, ill-defined mass was palpable in the right pelvis and lower abdomen. The mass was non-tender,
soft and non-mobile. The past history was nil particular. The routine blood and urine investigations were normal. An ultrasonography (USG) of the abdomen and kidney, ureter and bladder (KUB) was advised (Figure 1).

**Radiological Findings**

**Figure 1.** USG of abdomen/KUB – Elongated hypo to sonolucent mass in pelvis and abdomen; advice: CT scan (abdomen)

CT abdomen and pelvis

Based on the results of the USG report, a plain and contrast-enhanced CT scan of the abdomen and pelvis was advised (Figures 2 and 3).

The following investigations were carried out before the surgery:

- Blood Group: B Positive; Viral Markers: Negative; Amylase Level: 84 U/L; CRP: 1.2mg/L; TSH: 1.38 micro IU/ml; TC: 6800/cu mm; DC-P: 66.9%; HB: 16.1gm%; Platelets: 158,000/cu mm; LFT-WNL.
- CEA: 7.32 ng/ml
- Colonoscopy: Normal study up to caecum

A laparotomy revealed a large cystic mass (10 × 5 cm.) with a thick wall arising from the appendix with a 2 cm. clear base (from the mass). An appendectomy was performed and the abdomen was closed with a tube drain. There were no palpable lymphnodes in the mesoappendix and mesentery.

The frozen part of the appendectomy specimen was negative for malignancy. Hence, right hemicolecctomy was abandoned. The post-operative period was satisfactory with steady recovery. Oral fluids were administered on the second day of the post-operative period. After discharge, the patient was active and was tolerating a normal diet. He was able to evacuate his bowels and bladder normally. His condition at the time of discharge was stable.

**Discharge diagnosis:** Cystic mucinous neoplasm of the appendix

**Biopsy revealed mucinous cyst-adenoma of the appendix**

The patient was advised a review after eight days.
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DISCUSSION

Primary neoplasm of the appendix is found in 0.5–1.0% of the appendectomy specimens sent for histopathological examination and various classifications have been proposed for the same. With the exception of carcinoid tumors, most of the appendix neoplasms have been observed in the middle-aged and elderly. Most of the appendix tumors are seen as mucocoele in the USG and CT scan of the patients, 30–50% with signs and symptoms of:

- Contrast-enhanced CT of abdomen and pelvis
- Histopathology report
- Follow-up USG (3–6 months): (a) Renal calculi observed and (b) No recurrence of mass lesion
Acute appendicitis are evaluated using imaging techniques; these patients may have an asymptomatic palpable mass, intussusception or GI bleeding. The rupture of a malignant mucocoele is clinically presented as pseudomyxoma peritonei. Depending on the preoperative imaging findings, a surgeon can decide his surgical approach. Additional surgeries can be avoided.

In three-dimensional imaging techniques, mucinous neoplasms primarily show mucocoele. A plain abdomen radiograph may demonstrate a soft-tissue density, well-defined mass in the right lower abdomen and pelvis. Calcification increases specificity. Curvilinear mural calcification has been observed in 50% of the cases. In our case, a soft-tissue density lesion was seen in the right abdomen with faint punctate calcification on the plain radiograph.

The barium enema examination usually reveals that mucocceles generate smooth extrinsic compression over the medial aspect of the cecum. In our case, we had not performed the barium enema examination.

The size and morphologic features of the lesion cannot be assessed with colonoscopy. The colonoscopy was normal in our case.

USG, CT and magnetic resonance imaging (MRI) scans are useful for evaluating appendix neoplasms. USG shows a mucocoele from a mucinous neoplasm as an oblong or ovoid cystic mass with or without dystrophic mural calcification. The hypoechoic internal echoes are typical. In our case, an elongated, well-defined sonolucent mass of 10.2 × 3.4 cm. was present in the right mid-abdomen. There was no acoustic shadowing to suggest calcification. There was no evidence of septations or increased vascularity on the color Doppler results. Associated cystic ovarian tumors or tubo-ovarian abscesses had to be excluded.

The differential diagnosis of mucocoele is peri-appendiceal abscess, enteric duplication cyst, mesenteric cyst and hydrosalpinx.

A CT scan has more advantages than USG and MRI when it comes to diagnosing appendix neoplasms. The anatomic location is better defined by multiple-detector computed tomography with oral contrast. In our case, the mass was elongated in shape, retrocaecal in location and showed an attenuation of 40–50 HU, suggesting a fluid-filled mass. The appendix could not be visualized.

Multiple-detector computed tomography is more sensitive in detecting mural calcification compared to plain radiographs. In our case, minimal calcification was seen in the upper pole of the mass.

Secondary complications and malignancy are better evaluated with three-dimensional imaging. Soft-tissue thickening, irregularity of walls, and peri caecal stranding of fat are the non-specific findings. Fat stranding is seen in both malignancy and secondary inflammation. Intraluminal gas or an air-fluid level within a mucocoele suggests secondary infection. In our case, stranding of the surrounding fat was observed. Mild thickening of the walls and enhancement during the post-contrast study were observed. Calcified intra-luminal pearly spherules are usually observed on the plain X-rays and CT scan in myxoglobulosis. Our case had no such findings. Colonic intussusception, right ureteral obstruction and bladder compromises are rare clinical presentations of the mucocoele. They were not present in our case.

Mucinous adenocarcinoma may be clinically present with slowly increasing abdominal girth due to the rupture of the mass lesion. On imaging, the classical features are widespread heterogeneous peritoneal locules, displacement and distortion of bowel, and scalloping effect on solid organs. Intra-abdominal linear, punctate and septal calcification may be seen. The primary tumor of the appendix may still be apparent in a few cases. Our case had no such findings.

MRI delineates the cystic mass. It appears hypo-intense on T1 and hyper-intense on T2 sequences. However, calcification is poorly
delineated on the MRI. Contrast-enhanced MRI may show smooth mucosal enhancement in simple lesions. Adenomas show enhancing nodules and solid components. However, in our case, an MRI was not performed.

MANAGEMENT & PROGNOSIS
Surgical management depends on clinical presentation, extent of lesion, and histologic findings of the mucinous neoplasm. Retention cysts and small mucinous adenomas are surgically treated with simple appendectomy. The bulky adenomas with a large base may need caecal resection. In our case, the appendix base was free. The frozen section showed no malignant changes. There were no palpable lymph nodes in the mesoappendix and mesentery. An appendectomy was performed. Right hemicolectomy was abandoned.

The prognosis of adenomas and retention cysts is good, and the chances of a five-year survival are 91–100%. Adenomas of the appendix usually do not recur. Yantiss et al. found that 96% of the patients with a cellular extra-appendiceal mucin were disease-free. Misdraji et al. reported that patients with malignant peritoneal disease had a survival rate of 90% at three years and 44% at five years. Patients with low-grade peritoneal disease had a survival rate of 100% at three years and 86% at five years. Among those with mucinous adenocarcinoma, the chances of a 10-year survival were less than 10%. The features associated with poor prognosis include advanced stage and high-grade malignancy. In females, metastases to the ovaries are common. We did not find any recurrence of the mass lesion or metastasis during our six-month follow-up.

CONCLUSION
The diagnosis of cystoadenoma of the appendix is rarely considered before an elective surgery. Abdominal imaging is useful for preoperative diagnosis and for excluding the presence of peritoneal disease or other concomitant malignancies. An elevated CEA requires careful investigation to exclude malignancy. Appendiceal mucoceles more than 2 cm in size need surgical consultation. Proper communication between the radiologist, pathologist and surgeon is necessary for optimal patient management.

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REFERENCES

