Mucocele of the appendix: Incidental finding of a rare occurrence

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ABSTRACT

Mucocele of the appendix is a rare disease. Clinically, appendeceal mucocele (AM) is an incidental finding or it may mimic appendicitis. In AM, distension of the appendix is caused by mucus, the source of which can be benign or malignant. For the benign disease, appendectomy alone is sufficient but for malignant conditions adjunct treatments are needed. Proper pre-operative evaluation is imperative to distinguish between benign and malignant causes in order to guide the management and reduce complications. For the definitive diagnosis, histology and immunohistochemistry are required. We report an incidental finding of an appendeceal mucocele in a kidney donor during pre-transplant evaluation.

Introduction

Mucocele of the appendix is dilatation of appendix due to obstructive pathology caused by intra luminal mucoid material accumulation^{1,2}. Clinically, appendeceal mucocele (AM) is an incidental finding or it may mimic acute appendicitis (AA). Preoperative investigations help distinguish between AA and AM to decide the best surgical approach to prevent peritoneal spillage regardless of the etiology.

We report an incidental finding of an AM in a 55-year-old prospective kidney donor during her pre-transplant evaluation.

Case report

A 55-year-old female presented in surgical outpatient clinic. Patient was a would-be kidney donor to her daughter. Clinical examination during preoperative evaluation suggested a right iliac fossa mass. There was no significant past history relating to the mass. Rest of the systemic examinations were unremarkable. Provisional diagnosis of renal cyst or caecal tumor was made. Abdominal ultrasonography revealed an avascular tubular structure with calcified and echogenic debris and the tip abutting the lower pole of right kidney (9.4x4x3.5cm). Her biochemical investigations including CEA and CA19-9 were normal. Contrast enhanced CT (Fig. 1) was suggestive of AM.

On laparoscopic evaluation, a mass was seen originating from the appendix lateral to the caecum which was excised. The histopathological examination showed retention cyst of the appendix which was negative for malignancy^{2,3}. She had an uneventful recovery.

Discussion

Appendectomy is one of the most commonly performed gastrointestinal surgeries worldwide. AM is a relatively uncommon disease and may present as an incidental finding or is discovered on pathological examination⁴. The incidence of AM is 0.2 % to 0.4% in the entire appendectomy specimen³. It is of benign or malignant type which is differentiated histologically^{3,5}.

Rokitansky was the first to describe mucocele of appendix in 1942⁶. AM is described as appendix distended by mucus, due to any



Figure 1: a. Coronal CT, **b.** Axial CT scan showing the typical appearance of an appendeceal mucocele with cystic content and mural calcifications.

obstructing pathologies - mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%) and mucus retention cyst or lumen occlusion by carcinoid tumor⁷. With female preponderance, it is most common in the sixth or seventh decade of life⁸.

The common presenting symptoms of AM are episodic right lower quadrant abdominal pain, abdominal mass, weight loss and change in bowel habits. Complications include intussusception, bleeding, perforation, peritonitis and pseudo-myxoma peritonei^{9,10}.

CT is the investigation of choice. The typical CT appearance of AM is a well-circumscribed, smooth, thin walled mass with or without mural calcification located in the right iliac fossa. Regardless of the etiology, there may be variation in thickness of the wall of the mucocele and density depending on the contents. Most mucoceles have a low cystic watery density with possibility of high soft tissue attenuation. The most important feature is no exhibition of signs of periappendicular inflammation or collection^{8,11-13}. Typical mural calcification on a CT is seen only in half of the cases^{9,14,15}. Oval shaped cystic mass on ultrasound with acoustic shadow of mural calcification- "onion ring" is a typical finding. Like in CT, there is no sign of inflammation or periappendicular collection¹⁴⁻¹⁷. MRI is inferior in detecting calcifications compared to CT. Low sensitivity of FDG PET in detecting appendeceal carcinoma limits its use in workup of AM^{18, 19}.

Conclusion

Differentiating benign from malignant mucocele of appendix with modern radiological modalities is still a formidable challenge. Surgery with complete resection of the appendix is sufficient for retention cyst, cystoadenoma and mucosal hyperplasia. For cystadenocarcinoma, choice of treatment depends on the extent of the disease - if no mesenteric or other organ involvement is seen then resection is enough for complete cure²⁰.

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