

Case Report

Appendicular Mucocele - A Case Report

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Kuwait Medical Journal 2008, 40 (1): 78-80

ABSTRACT

Appendicular mucocele by definition is a cystic dilatation of the appendiceal lumen by mucin accumulation. This is a rare lesion; its prevalence in appendectomy specimens being only 0.2 - 0.3%. It may be an outcome of various processes. Most important from the surgical point of view is the mucocele caused by mucinous cystadenoma and cystadenocarcinoma. Less commonly it could be

due to appendicolith, carcinoid *etc.* It is known to be associated with pseudomyxoma peritonei resulting from a rupture. It is therefore important to identify the disease process preoperatively and to plan a careful resection. We report here one case with surgical and histopathological confirmation.

KEY WORDS: appendix, imaging, mucocele

INTRODUCTION

Appendicular mucocele is a rare lesion. It is a descriptive term denoting an obstructive dilatation of the appendicular lumen by mucinous secretions. Mucinous cystadenoma and cystadenocarcinoma account for 60 - 70% of all mucoceles. Less common causes are retention cyst, mucosal hyperplasia, carcinoid, appendicolith, endometriosis, adhesions and volvulus. The clinical presentation is usually non-specific with 50% of cases being an incidental finding at surgery. Symptoms could be an indeterminate abdominal pain or chronic or intermittent abdominal colicky pain due to intussusception of the mucocele. Occasionally the patient could present with sepsis due to a superimposed infection.

CASE REPORT

A 26-year-old male presented with a mass in the right lower quadrant of the abdomen. He had felt the mass two months prior to presentation and this mass had been gradually increasing in size. The patient had a febrile illness at the onset of his symptoms, which had now abated. There was no change in bowel habits. On examination, the clinician felt a firm mass in the right iliac fossa, which was also mildly tender and mobile.

The patient's blood counts were normal. Plain abdominal radiograph showed a curvilinear calcification in the right side of the pelvis (Fig. 1). An ultrasound examination showed a 6.2 cm x 3 cm cystic mass with internal echoes and a mural

nodule arising from its wall (Figs. 2a & 2b). Pre and post contrast CT scan showed that this was a well encapsulated ovoid mass, medial to the cecum and extending inferiorly, with curvilinear calcification in a portion of its wall (Fig. 3). The wall was noted to enhance after contrast administration. A mural enhancing nodule was also identified (Fig. 4). No enhancement of the cystic contents was noted.

A radiological diagnosis of mucocele of the appendix was made. At surgery, the diagnosis was confirmed and the appendix was carefully resected (Fig. 5). The cecum, terminal ileum and mesentery were noted to be normal, and no evidence of infection was noted. The histopathological diagnosis was mucocele of the appendix on top of mucinous cystadenoma with microscopy showing atypical proliferating mucous cells and papillary formation with some areas showing mucosal atrophy with fibrosis and inflammation.

DISCUSSION

Mucocele of the appendix are rare lesions representing 0.2 - 0.3% of surgical appendectomy specimens. They are pathologically divided into four categories. A very rare type is secondary to occlusion of the lumen from post-inflammatory scarring, age related atrophy, congenital obstruction of Gerlach's valve or extramural compression. This type leads to an atrophic mucosa. All other types are classified into a spectrum, from mucous hyperplasia to mucinous cystadenoma to mucinous cystadenocarcinoma,

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Fig. 1: Plain radiograph showing curvilinear calcification on the right side of the pelvis

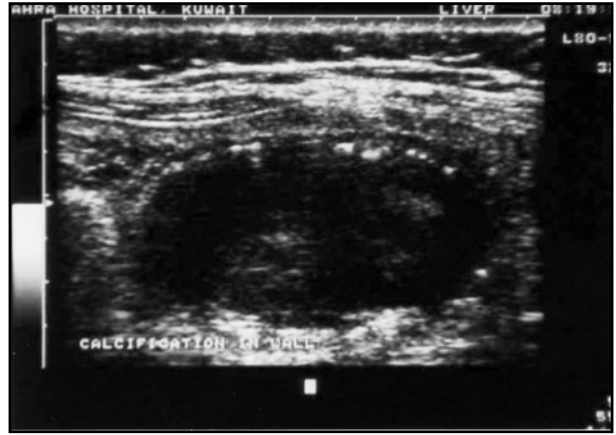


Fig. 2a: Ultrasound showing cystic mass with internal echoes and calcification in its wall

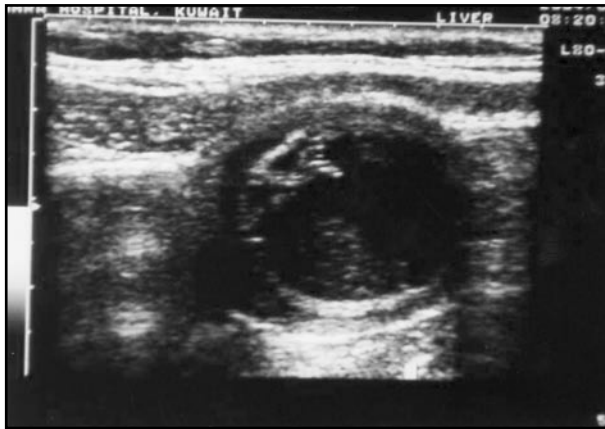


Fig. 2b: Mural nodule in wall of cystic mass

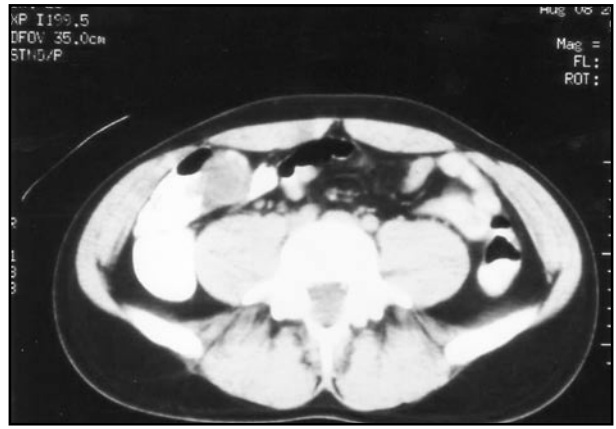


Fig. 3: Pre contrast CT showing discrete ovoid mass medial to the cecum with curvilinear calcification in its wall



Fig. 4: Post contrast CT showing enhancing wall and mural nodule

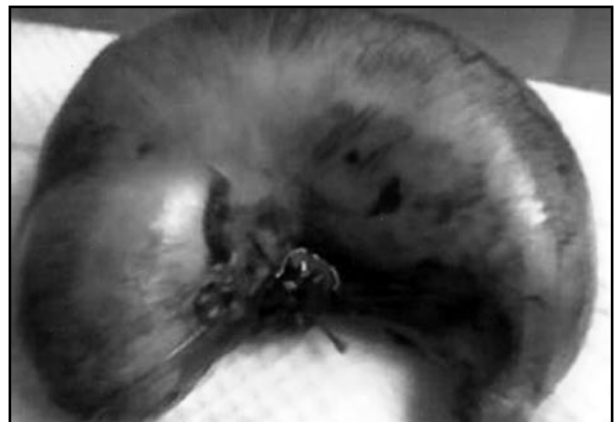


Fig. 5: Resected appendicular mucocele

depending on the pathology of the mucosa. Whatever may be the cause, obstruction of the lumen and accumulation of yellow mucous within the appendiceal lumen results. About 25% of mucoceles are from mucosal hyperplasia. These typically have minimal distension. Mucinous cystadenomas, which account for about 60% of mucoceles, are more markedly distended. However, they are typically asymptomatic, and found incidentally. Mucoceles up to 40 cm x 24 cm x 20 cm have been reported. About 20% have extra appendiceal

extrusion of mucus. If no cells are present in the peritoneal mucous, the prognosis is excellent. Mucinous cystadenocarcinomas, which constitute 10.15% of cases, are more likely symptomatic. This diagnosis is made by either neoplastic glands invading the wall or by the presence of cells in the peritoneal mucous. It is thought by some authors that pseudomyxoma peritonei is a complication of mucinous cystadenocarcinoma only.

Mucinous cystadenocarcinomas are extremely rare (benign: malignant about 10:1), but are

believed to arise in cystadenomas, and there is a high correlation of synchronous or metachronous colorectal adenomas and carcinomas (up to 20% in two series). There have also been reports of association with gastrointestinal tract, ovarian and kidney tumors. It is thought that only mucinous cystadenocarcinomas lead to pseudomyxoma peritonei^[1]. However, other authors believe this can complicate either benign or malignant mucoceles, although pseudomyxoma peritonei from the former would carry a better prognosis^[2-4]. A very important fact to be stressed here is the need for more mucoceles of the appendix to be diagnosed preoperatively. This makes the surgeon aware of the need for more careful surgery and consequently reduces the chances of iatrogenic damage to a mucocele with resultant leakage of the contents in the abdominal cavity with serious repercussions, especially pseudomyxoma peritonei^[1].

Onsonography, there is typically excellent through transmission and posterior wall enhancement. When the wall is calcified, posterior acoustic shadowing may occur, but often cannot be appreciated. The wall thickness varies, but if the wall is greater than 6 mm, one should also consider uncomplicated acute appendicitis. The internal features vary from anechoic to hyperechoic, and may be dependent. Internal septations, polypoid lesions extending into the lumen and irregular outline seem to be associated with the malignant variety, although some papillary processes may be seen in mucinous cystadenomas^[1,5,6]. The differential diagnosis on ultrasound include fluid filled small bowel, fluid in a small or large bowel diverticulum, appendiceal / diverticular abscess, mesenteric cyst seroma and particularly in females of reproductive age group, salpingitis and ectopic pregnancy masses^[5,7].

On CT, typically it is a low-attenuation (0 - 40 H.U.) smooth or lobulated mass. The more complex and irregularly shaped mucoceles tend to be mucinous cystadenocarcinomas. They may have single or multiple cystic components and some solid component. There may even be infiltration into adjacent organs such as the colon, ureter and bladder. Curvilinear or punctuate calcification in the lesion is

strongly suggestive of mucinous cystadenoma, and this is often not seen on plain films. Amorphous calcifications may be seen in the malignant type. This is from chronic inflammatory process incited by the irritating mucous. Vertical folds, mimicking intussusception, have also been described. A pitfall is that the fluid filled terminal ileum may resemble a mucocele, so delayed scanning may be warranted in some cases^[5,6].

The finding of an appendiceal mucocele should prompt a search for an associated tumor as there is six-fold increased incidence of colon adenocarcinoma and there may be association with mucin-secreting tumors of the ovary^[1].

CONCLUSION

Appendicular mucocele is to be considered in the differential diagnosis of a right iliac fossa mass and CT scan is imperative in the correct preoperative diagnosis. This helps the surgeon to be more careful and it reduces the risk of iatrogenic rupture of the mucocele with resultant leakage of its contents into the abdominal cavity causing pseudomyxoma peritonei^[7].

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