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CASE REPORT

Mucocele of the Appendix with Hematuria

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Mucocele of the appendix, a nonspecific and descriptive term for an abnormal mucous accumulation within the appendiceal lumen, regardless of the underlying cause, is a rare clinical disease that is not usual as a consideration in the differential diagnosis of right lower quadrant lesions. The reported prevalence of mucocele of the appendix in appendectomy specimens is 0.2-0.3 percent. Hematuria due to mucocele of the appendix is extremely rare. Up to now, only few cases of such condition have been recorded in the literature. We describe a woman who experienced intermittent episodes of right lower quadrant pain and hematuria. Abdominal exploration incidentally displayed mucocele of the appendix. No evidence of other lesions was found. The patient was still in good health after operation. This case highlights mucocele of the appendix as a consideration in the differential diagnosis of right lower quadrant pain with hematuria.

INTRODUCTION

Mucocele of the appendix, the gross enlargement of the appendix caused by accumulation of mucus within the appendix, is a rare lesion [1]. It is encountered in only 0.2 to 0.3 percent of 43,000 appendectomies [1]. It may be caused by neoplastic or non-neoplastic lesions [2]. Mucocele of the appendix can present like most appendiceal pathology with either mild abdominal pain or life-threatening peritonitis [2]. The four histologic subtypes include retension cysts, mucosal hyperplasia, cystadenomas, and cystadenocarcinomas, with relative frequencies of 18, 20, 52, and 10 percent, respectively [3, 4]. The course and the prognosis of mucocele of the appendix are related to these subtypes. We report one rare case with an unusual presentation of hematuria caused by mucocele of the appendix. We also review the appropriate literature about the clinical, radiologic, and diagnostic characteristics of this rare entity.

CASE REPORT

A 67-year-old woman was admitted to our hospital because intermittent episodes of right lower quadrant pain and microhematuria, which was observed at one local hospital three days before admission. Physical examination revealed no lymphadenopathy. No enlargement of liver or

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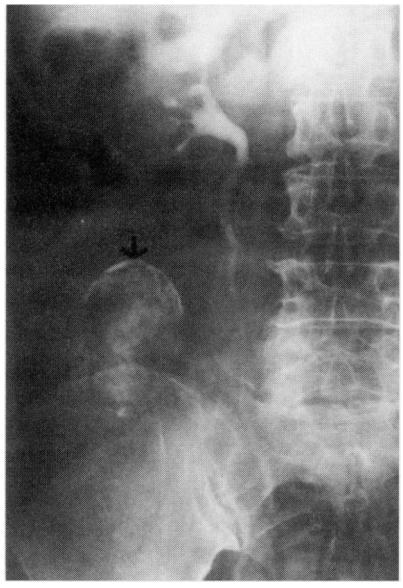

Figure 1. Intravenous pyelography shows one lesion with eggshell calcified wall in right lower quadrant (arrow).

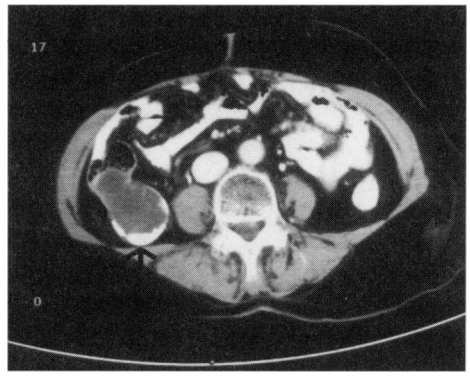

Figure 2. Abdominal CT scan shows a cystic well-encapsulated lesion with curvilinear calcified wall in right lower quadrant and indenting the cecum

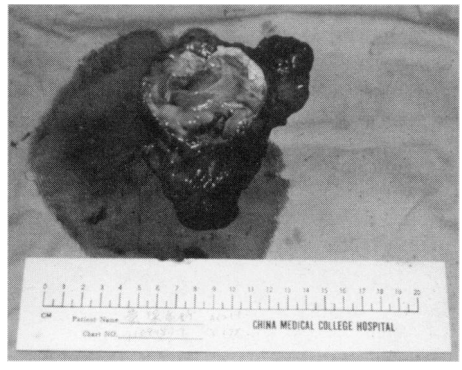

Figure 3. Opened surgical specimen shows mucinous material with calcified wall.

spleen was noted. No palpable mass or rebound pain was exhibited in the right lower quadrant, but local tenderness was noted in the right lower quadrant. No pain was noted in the right flank area in response to percussion. She denied any history of urolithiasis. After admission, laboratory data showed white blood cell count: 4.84 x 10^9/l. Urine analysis showed RBC: 8-10/high power field and WBC: 0-1/ high power field.

The intravenous pyelography showed one lesion with an eggshell calcified wall in the right lower quadrant (Figure 1). No evidence of filling defect or calcification in the urinary tract was observed. The abdominal computed tomography (CT) scan showed one cystic well-encapsulated lesion with a curvilinear calcified wall in the right lower quadrant (Figure 2). Therefore, under the impression of mucocele of the appendix, abdominal exploration was performed. During operation, one well-encapsulated mass of 5.0 x 5.0 x 5.0 cm in size was found in the retrocecal area. The mass was adherent to adjacent ileum and cecum. Thus, right hemicolonectomy was performed smoothly.

The opened surgical specimen showed mucous material with calcified wall (Figure 3). Pathologic examination showed a mucinous cystadenoma. The cavity was lined with proliferating epithelium and mild atypical changes were also noted.

DISCUSSION

In a review of 60 cases with mucocele of the appendix by Aho et al. [3], the male: female ratio was 1:4, with a mean age of 55 years. Clinically, the most common presentation was right lower quadrant pain. Other reported symptoms included intermittent colic pain caused by intussusception of the mucocele into the cecum, gastrointestinal bleeding associated with intussusception, and a palpable abdominal mass. Leukocytosis was usually absent, unless the mucocele was secondarily infected [1-3]. Of all mucoceles, 23 to 50 percent were incidental findings during laparotomy for a suspected appendicitis or during abdominal exploration for other reasons [4, 5]. A early, correct preoperative diagnosis of mucocele of the appendix based on abdominal pain, a palpable mass in the right lower quadrant, or conventional radiography was rare [6], because these clinical symptoms were often nonspecific, vague, or absent. However, it is important for adequate diagnosis and resection because of the risk of rupture and development of *pseudomyxoma peritonei*, a potentially fatal entity [7].

CT scan and sonography have made possible the early detection of mucocele of the appendix. On CT scan, the presence of a curvilinear or punctate cystic lesion with calcified wall in the right lower quadrant strongly indicates mucocele of the appendix [8]. On sonography, the diagnosis of mucocele of the appendix is strongly suggested if a cystic mass with calcified wall and epithelial lining or papillary processes is demonstrated in the right lower quadrant in a patient without appendectomy [9]. If mucocele of the appendix is identified preoperatively, it is recommended that it should be removed because there might be an unrecognized cystadenocarcinoma of the appendix [10].

Mucocele of the appendix rarely exhibits urologic features. In Baskin and Stoller's report [11], two cases presented with pelvic masses causing urinary frequency, and one case with fever and hydronephrosis. In the previous reports [2, 12, 13], hematuria was a rare urologic sign. These presentations might be caused by local or mass effects of mucocele of the appendix [2]. In our case, hematuria was a rare finding, which might be due to ureteric compression by the mucocele. Because leukocytosis was not revealed, the mucocele was not secondarily infected. Although intravenous pyelography showed one lesion with eggshell calcified wall in right lower quadrant, it was still difficult to correctly diagnose mucocele of the appendix based on clinical features preoperatively. Therefore, use of sonography or CT scan made possible early detection of mucocele of the appendix. As the literature reveals, laparoscopy can also be used as a diagnostic tool in equivocal cases, or for an appendectomy [2].

In conclusion, although rarely found, urologic signs and symptoms may be a feature of appendiceal pathology. This case illustrates the importance of careful assessment of right lower quadrant pain with hematuria caused by the unusual appendiceal pathology when a urinary tract lesion was not found.

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