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CASE REPORT

Porcelain Appendix: A Rare but Essential Diagnosis

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ABSTRACT

Mucocele of the appendix is a rare entity, not often considered when problems pertaining to the right lower abdominal quadrant are encountered. We describe the case of a patient with a large cystic mass in the lower abdomen, with extension into the pelvis. Pathological examination confirmed the diagnosis of appendiceal mucocele due to underlying cystadenoma. The condition of appendiceal mucocele is discussed and diagnostic imaging features described.

Key Words: *Appendix, Diagnostic imaging, Mucocele*

CASE REPORT

A 61-year-old man with a known history of hyperlipidaemia and ischaemic heart disease was admitted with rectal bleeding. On physical examination, the abdomen was soft and non-tender. Normal bowel sounds were present. However, a large right lower quadrant mass was noted. Ultrasound demonstrated a large circumscribed cystic lesion in the right lower quadrant extending to the pelvis (Figure 1). The lesion was extrinsic to the urinary bladder and kidney, and contained echogenic internal debris. The adjacent bowel loops were displaced but invasion was not seen. There was no local tenderness on graded compression and no free peritoneal fluid was evident. The lesion appeared to arise from the ascending colon, caecum, or appendix.

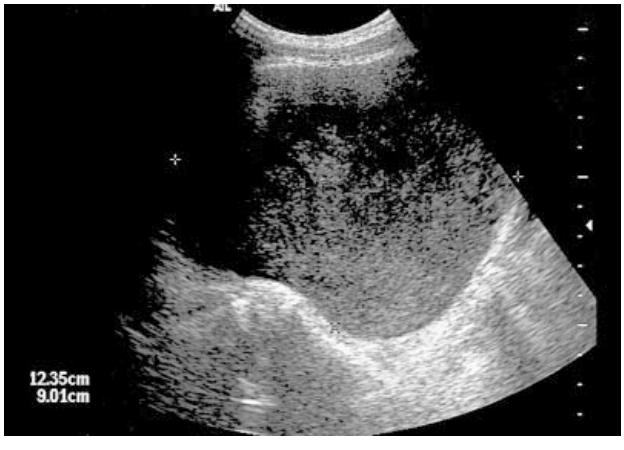

Figure 1. Ultrasonography of the abdomen demonstrated a well-defined cystic mass in the right lower quadrant with internal debris. The lesion had a thin wall with no evidence of acute inflammation.

In view of the clinical presentation of per rectum bleeding, colonoscopy was performed. No abnormal intraluminal lesion could be located as far as the caecum. Subsequent contrasted-enhanced computed tomography of the abdomen (Figure 2) depicted a large cystic mass in the right lower quadrant with extension to the pelvic cavity. The lesion had an incomplete calcified rim and minimal rim contrast enhancement was noted. A preoperative diagnosis of appendiceal mucocele was made.

Laparotomy confirmed the presence of an 11.5 cm x 18 cm cystic lesion originating from the vermiform appendix. Microscopic examination of the surgical specimen revealed an underlying mucinous cystadenoma of the appendix. The patient's postoperative course was uneventful.

DISCUSSION

Appendiceal mucocele is a descriptive term for abnormal mucus accumulation causing distension of the appendiceal lumen, irrespective of the underlying cause.[1] It is a rare clinical entity. The reported prevalence in appendectomy specimens at surgery is 0.2-0.3%.[2] The underlying pathology can be classified

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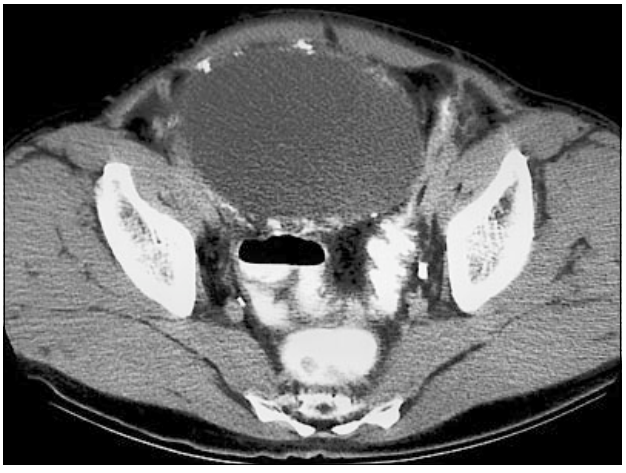

Figure 2. Postcontrast contiguous CT scanning depicted a well-defined homogeneous cystic lesion in the lower abdomen. Rim calcifications similar to calcifications found in a porcelain gallbladder were seen. No abnormal diffuse intraperitoneal lesion was apparent.

into four histological subtypes: retention cyst, mucosal hyperplasia, cystadenoma, and cystadenocarcinoma.[3-6] The latter two are mucin-secreting appendiceal tumours accounting for 62-76% of all cases of appendiceal mucocele, with cystadenoma seen in the majority of these cases.[3,6] Previously, it was thought that most mucoceles occurred as a result of postinflammatory obstruction or secondary to an obstructive faecaelith with distal accumulation of mucus.

Appendiceal mucocele usually presents with non-specific symptoms and up to 50% of cases are found incidentally at surgery.[5,7] Symptoms related to mucocele include vague abdominal distress and pain, chronic or intermittent colicky pain caused by intussusception of the mucocele, a right iliac fossa mass, sepsis, or urinary symptoms. Aho et al[3] reported that almost a quarter of cases were asymptomatic and discovered incidentally. In this patient, the presenting clinical symptom was per rectum bleeding while the lesion was found only incidentally on clinical examination. Often the diagnosis cannot be made preoperatively on the basis of the clinical examination alone; plain film radiography is also usually not helpful. Accurate diagnosis is important, however, because of the risk of rupture at surgery, the potential development of pseudomyxoma peritonei, and the possibility of malignant transformation.[8] When pseudomyxoma peritonei is found in the setting of appendiceal cystadenocarcinoma, the prognosis is poor, with a 5-year survival rate of 20%.[2]

The typical finding in appendiceal mucocele is of a circumscribed cystic mass in the right lower abdominal quadrant. Plain film radiographs may reveal mass effect on the adjacent bowel or urinary bladder. Ultrasonography may depict an oblong or rounded (when a very large mass) heterogeneous cystic mass containing internal echoes because of mucin content. The degree of internal echogenicity is related to the number of acoustic interfaces provided by the mucin.[9] Through-transmission and posterior enhancement are usually present. However, one report described strand-like layers of varying echotexture suggesting a solid lesion.[10] The mass is usually well-encapsulated unless rupture has occurred. The primary sonographic feature differentiating appendiceal mucocele from uncomplicated acute appendicitis is a lack of appendiceal wall thickening (i.e. wall thickness <6 mm compared to >6 mm in acute appendicitis).

CT scan demonstrates a low-attenuated, well-encapsulated mass in the right lower quadrant, with a smooth regular wall. If the lesion is large enough to have midline or pelvic extension, it can be difficult to delineate the origin of the lesion. The degree of attenuation depends on the amount of mucin in the mass. Adjacent bowel is displaced but there is no related periappendiceal inflammation or abscess, which is an important differentiator of this condition from acute appendicitis. Thorough opacification and distension of the bowel lumen by oral contrast is essential to exclude the possibility of a lesion arising from the surrounding bowel loops. The presence of curvilinear or punctate wall calcification in the right lower quadrant strongly suggests the diagnosis of appendiceal mucocele.[8] Calcifications are thought to be a dystrophic response to a chronic inflammatory process incited by mucus in the appendiceal wall, ultimately leading to the development of a "porcelain" appendix.[2] The presence of wall calcification in a right lower quadrant cystic mass with intraluminal gas allows one to differentiate an infected mucocele from an abdominal abscess cavity.

Magnetic resonance imaging findings of an appendiceal tip mucocele have been reported. Intermediate signal intensity on T1-weighted images and homogeneous high-signal intensity on T2-weighted images were seen, reflecting the high protein content of the lesion.[11] Of note, MRI appears less effective than CT scanning for the detection of mural calcifications.

The differential diagnosis of appendiceal mucocele includes mesenteric cyst and duplication cyst. Enteric duplication is rare in adults and calcification is rare in

mesenteric cysts. A right ovarian cyst or hydrosalpinx should also be considered in the differential diagnosis in women. In a patient who has not had an appendectomy, CT findings of a cystic mass containing mural calcification in the anatomic location of the appendix and adherent to the adjacent caecum are characteristic of an appendiceal mucocele, however.[12]

CONCLUSION

We present the case of a patient with a lower abdominal cystic mass and rim calcifications. Porcelain appendix arising from mucinous cystadenoma (carcinoma) should be considered in the differential diagnosis of such cases in order to avoid the risk of rupture at surgery with subsequent development of pseudomyxoma peritonei.

REFERENCES

1. Lo RH, Kan PS. Appendiceal mucinous cystadenoma presenting as "porcelain" appendix with myxoglobulosis—a rare cause of a right lower quadrant mass. Singapore Med J 1998;39: 174-176.
2. Dachman A, Lichtenstein J, Friedman A. Mucocele of the appendix and pseudomyxoma peritonei. AJR Am J Roentgenol 1985;144:923-929.
3. Aho AJ, Heinonen R, Lauren P. Benign and malignant mucocele of the appendix. Histological types and prognosis. Acta Chir Scand 1973;139:392-400.
4. Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal "mucocele". Cancer 1973;32:1525-1541.
5. Wolff M, Ahmed N. Epithelial neoplasms of the vermiform appendix. Cancer 1976; 37:2511-2521.
6. Landen S, Bertrand C, Maddern GJ, et al. Appendiceal mucocele and pseudomyxoma peritonei. Surg Gynecol Obstet 1992;175: 401-404.
7. Isaacs KL, Warshauer DM. Mucocele of the appendix: Computed tomographic, endoscopic and pathologic correlation. Am J Gastroenterol 1992;87:787-789.
8. Madwed D, Mindelzun R, Jeffrey RB. Mucocele of the appendix: imaging findings. AJR Am J Roentgenol 1992;159:69-72.
9. Horgan J, Chow P, Richter JO, Rosenfield AT, Taylor KJ. CT and sonography in the recognition of mucocele of the appendix. AJR Am J Roentgenol 1984;143:959-962.
10. Gibbs M. Mucinous cystadenoma and cystadenocarcinoma of the vermiform appendix with particular reference to mucocele and pseudomyxoma peritonei. J Clin Pathol 1973;26:413-421.
11. Takahashi S, Furukawa T, Ueda J. Case report: Mucocele of the tip of the appendix. Clin Radiol 1998;53:149-150.
12. Zissin R, Gayer G, Kots E, Apter S, Peri M, Shapiro-Feinberg M. Imaging of mucocele of the appendix with emphasis on the CT findings: a report of 10 cases. Clin Radiol 1999;54: 826-832.