Singapore Med J 1998; Vol 39(4):174-176

Appendiceal Mucinous Cystadenoma Presenting as "Porcelain" Appendix with Myxoglobulosis – A Rare Cause of a Right Lower Quadrant Mass

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ABSTRACT

Mucinous cystadenoma is a rare tumour of the vermiform appendix and is associated with cystic dilatation of the appendix, to which the more general term of mucocoele has been applied. Mucocoele of the appendix is only a descriptive term for abnormal mucus accumulation causing distension of the appendiceal lumen, irrespective of the underlying cause. Pre-operative diagnosis of appendix mucocoele, though infrequently made, is important as some of these lesions may be malignant, and also is essential in order to avoid the risk of rupture at surgery with subsequent development of pseudomyxoma peritonei. The appearances of mucocoele of the appendix and its uncommon variant of myxoglobulosis on plain radiograph, ultrasound and barium study are presented, together with review of the literature.

Keywords: mucinous cystadenoma, vermiform appendix, appendix mucocoele, pseudomyxoma peritonei, myxoglobulosis

CASE REPORT

A 61-year-old Chinese male presented with a 3-week history of fresh bleeding per rectum, weight loss and occasional abdominal pain. Apart from a history of prolapsed haemorrhoids and a similar episode of bleeding per rectum the previous year, which subsided spontaneously without treatment, he had no major medical illness or surgery previously.

Physical examination of the abdomen revealed a smooth tubular mass in the right iliac fossa which was initially thought to be faecal laden caecum. Haemorrhoids were noted on rectal examination and ligated on proctoscopy. Sigmoidoscopy was normal up to 15 cm.

Initial laboratory investigations revealed only a slightly reduced haemoglobin level (10.8 g/dL), all other parameters being within normal limits.

The patient was followed up in outpatient clinic two months later. By then the rectal bleeding had ceased but the right iliac fossa mass was still present. Colonoscopy was subsequently performed and was normal up to the hepatic flexure, but unable to proceed further which was suspected to be due to distorted anatomy by the right lower quadrant mass. Abdominal ultrasound examination revealed a dilated, cystic tubular mass filled with echogenic debris with some acoustic shadowing in the right iliac fossa, as well as associated wall calcification with distal acoustic shadowing (Fig 1a and 1b). A barium enema was also requested; the preliminary plain film of which showed a tubular mass in the right iliac fossa with curvilinear wall calcification as well as clusters of small rounded calcific densities within (Fig 2). The calcified mass was shown to be extra-colonic in origin, located postero-inferior to the caecum, and causing mild indentation on the caecal pole (Fig 3). Normal appendix was not visualised or identified in the enema study. Incidental colonic diverticulae and right renal calculi were also noted.

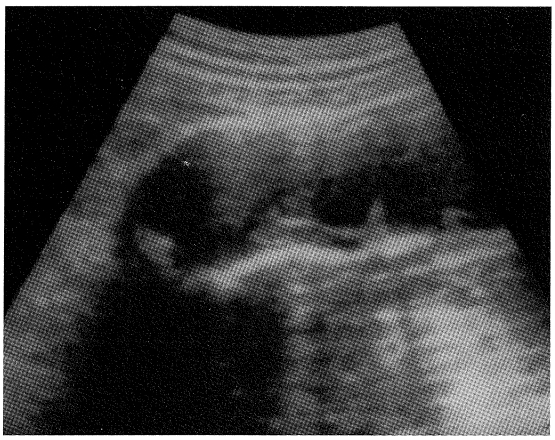

Fig 1a – Longitudinal ultrasound scan of the right lower quadrant mass. Mass has cystic appearance with tubular shape and is filled with internal echogenic debris. Associated wall calcification with distal acoustic shadowing is also noted to be present.

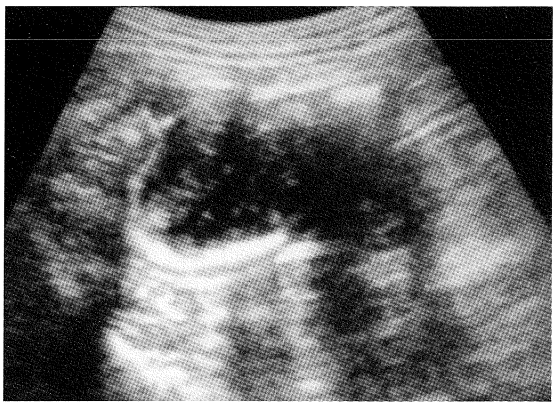

Fig 1b – Transverse ultrasound scan through the same mass. Note again its cystic appearance and presence of wall calcification with distal acoustic shadowing.

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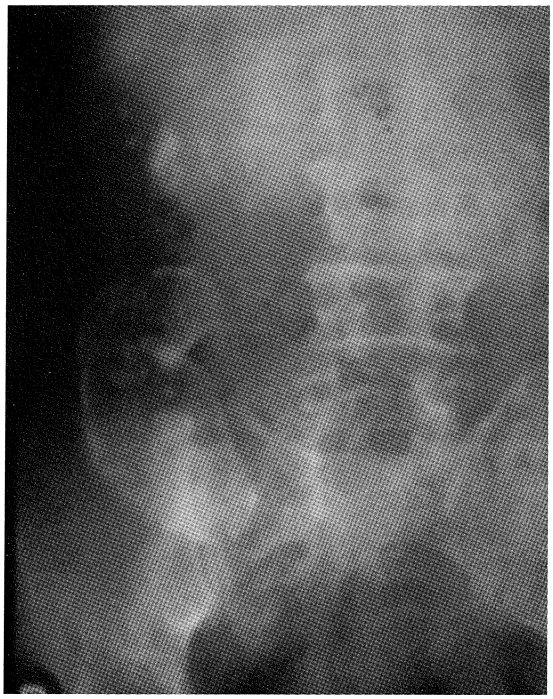

Fig 2 – Plain abdominal radiograph. Note the tubular mass in the right lower quadrant with curvilinear wall calcification and some clusters of small rounded calcific densities within. Incidental right renal calculi are also present.

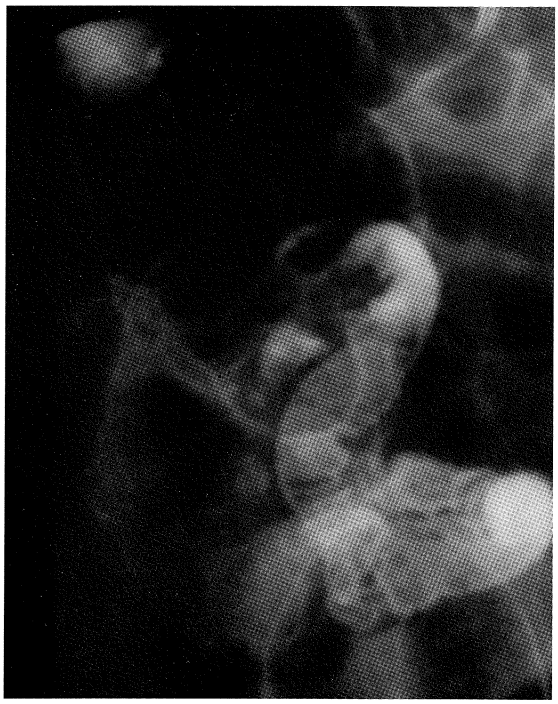

Fig 3 – Barium enema study. Localised oblique view of caecal region revealing the mass lesion to be extra-colonic in origin and located posterior and inferior to the caecal pole. Reflux of barium into the terminal ileum is demonstrated. Normal appendix is not identified.

Based on imaging features, the differential diagnoses therefore included mucocoele of the appendix, a pericolic inflammatory mass or abscess, possibly secondary to diverticulitis, duplication cyst, mesenteric cyst and haematoma. The patient subsequently underwent exploratory laparotomy which revealed an 8 x 4 cm paracolic appendiceal cystic mass containing jelly-like mucoid material. Appendicectomy was performed and on histology, a diagnosis of mucinous cystadenoma of the appendix was made.

DISCUSSION

Mucocoele of the vermiform appendix is simply a descriptive term for dilatation of the appendiceal lumen due to an abnormal accumulation of mucus, regardless of the underlying cause. It is a relatively rare entity, the reported incidence being 0.2% to 0.3% of appendicectomy specimens at surgery[1-3]. Recent evidence from pathological examinations suggests that mucosal atypia or neoplasia may play a role in mucocoele formation[4,5]. Current classification of appendix mucocoele include the following major categories: (1) retention mucocoele with normal appendiceal mucosa or inflammatory granulation tissue due to simple obstruction of the lumen, frequently with secondary infection; (2) focal or diffuse mucosal hyperplasia; (3) mucinous cystadenoma mucocoele with neoplastic epithelium similar to that in villous adenomas and adenomatous polyps of the colon, and (4) mucinous cystadenocarcinoma mucocoele with neoplastic epithelium similar to that in adenocarcinomas of the colon. In a study on appendiceal neoplasms, Wolff and Ahmed[5] found a significant association of adenomas of the appendix with other neoplasms, especially gastrointestinal neoplasms. 21.4% of the patients in their series had colonic carcinoma, while 23.8% had one or more colonic neoplasms.

Over the past decades, the diagnosis of mucocoele of the appendix has been made at laparotomy for suspected appendicitis[6], and pre-operative diagnosis has rarely been made[3,6]. More recently, however, modern, non-invasive imaging modalities especially ultrasound and CT, have been shown to be extremely useful in the pre-operative diagnosis of appendix mucocoele[1,7], as well as demonstrating the presence of pseudomyxoma peritonei which may develop as a complication of rupture of the mucocoele[7-9].

On a plain abdominal radiograph, an appendix mucocoele may be evident as a soft tissue mass in the right lower quadrant, with mass effect on the adjacent loops of bowel or the bladder. Curvilinear or punctate wall calcification may be present, as clearly demonstrated in the case presented (Fig 2), and is thought to be due to a dystrophic response to chronic inflammation which can ultimately result in a "porcelain" appendix. The mucocoele usually causes some indentation on or displacement of the caecum (or adjacent terminal ileum or sigmoid colon) without ulceration, and barium enema findings are those of a submucosal or extrinsic mass with intact overlying mucosa and non-filling of the appendix. A "star" pattern to the mucosa at the base of the appendix has been observed, and may represent caecal mucosal folds radiating toward the obstructed appendiceal orifice, as well as a "vortical fold" pattern representing a concentric ring appearance of caecal mucosal folds directed toward the obstructed appendiceal orifice[2]. Both patterns, however, are reported to be non-specific.

Ultrasound examination may reveal a heterogeneous cystic mass extrinsic to solid abdominal viscera. It appears well-encapsulated unless the mucocoele has ruptured, and this is usually through-transmission with posterior enhancement. However, a highly echo-dense wall with distal acoustic shadowing may indicate calcification, as was observed with the case presented here (Figs 1a and 1b). The main feature on ultrasound distinguishing it from uncomplicated acute appendicitis is the absence of thickening of the appendiceal wall greater than 6 mm[7], the latter of which would appear as a characteristic target lesion (an echogenic submucosal layer lying in between hypoechoic oedematous inner lamina propria-muscularis mucosa and outer muscular layer).

CT features include a hypoattenuated, well-encapsulated, right lower quadrant mass with smooth margins. The density of the mucocoele depends on the mucin content within, ranging from near water density to soft tissue density. There may be mass effect with displacement of the adjacent bowel loops, but no significant peri-appendiceal inflammatory changes or abscess formation, features which help to differentiate it from acute appendicitis. CT will also readily demonstrate the presence of wall calcification.

A variation in the appearance of appendix mucocoele occurs in myxoglobulosis when the appendix becomes filled with "translucent globules" or a cluster of "frog eggs", with a reported incidence of 0.35% – 8% of cases[2]. These globules become evident on plain films, ultrasound and CT scans when they become calcified as demonstrated in this case presentation (Figs 1a, 1b, 2), and may shift within the mucocoele. The appearance is considered pathognomonic when the calcification can be localised to the appendix. It has been suggested that mucin within the mucocoele is "organised" by the extension of granulation tissue from the wall of the mucocoele which subsequently breaks off and undergoes necrosis and calcification[2].

Complications may arise from formation of an appendix mucocoele. Intussusception of a mucocoele is rare, with less than 30 cases reported in the literature. The mucocoele acts as an intussusceptum and becomes invaginated into the caecum, producing the appearance of an entero-enteric intussusception which, on barium studies, may be evident as a "coiled spring" appearance. CT appearance may show a collapsed proximal segment surrounded by a rim of mesenteric fat lying within the distal lumen[7].

Another possible complication is pseudomyxoma peritonei which occurs when the mucocoele ruptures into the peritoneum, releasing the mucinous contents into the abdominal cavity[9], with resultant implants of mucinous epithelium on the peritoneal surfaces and mucus accumulation within the peritoneal cavity. It is readily demonstrated on CT scan which shows low-attenuation ascites with scalloping of the liver and/or splenic contour, corresponding to the peritoneal implants. Adhesions and intestinal obstruction may result. It has also been suggested that pseudomyxoma peritonei occurs only in the presence of a "malignant" mucocoele[10], although recent studies do not appear to support this[2]. However, when it occurs in the presence of appendiceal cystadenocarcinoma, the prognosis is grave with only a 20% 5-year survival rate[7].

In conclusion, the imaging findings of an appendix mucocoele include the following features: a right lower quadrant soft tissue mass which may show curvilinear or punctate wall calcification on the plain radiograph; an extrinsic impression on the caecum, terminal ileum or sigmoid colon with "vortical fold" or "star" pattern of the caecal mucosa in a barium study; a heterogeneous cystic tubular mass on ultrasound with variable sonographic echogenicity secondary to anechoic fluid and echogenic mucus but excellent through-transmission unless calcified in which case there may be distal acoustic shadowing; and on CT scan, a low-attenuation, well-encapsulated mass with or without wall calcification. These combined features of a right lower quadrant mass in a patient who has not had a previous appendicectomy should alert the radiologist to the diagnosis of an appendix mucocoele as well as its complications such as pseudomyxoma peritonei which is most readily recognised on CT scan.

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