Archives of Gastroenterology and Hepatology
ISSN: 2639-1813
Volume 1, Issue 2, 2018, PP: 1-4

Appendiceal Mucocele

Francisco Miguel González Valverde[1]*, Patricia Pastor Pérez[2]

[1]Associate Professor. Department of Surgery. University of Murcia, Spain.
[2]Department of Surgery, Pediatrics, Obstetrics and Gynecology, Reina Sofía General University Hospital, Murcia, Spain.
migova67@gmail.com

***Corresponding Author:** Francisco Miguel González Valverde, MD, Associate Professor. Department of Surgery. University of Murcia, Spain.*

Abstract

Mucocele of the vermiform appendix is characterized by a dilatation of the appendiceal lumen due to abnormal accumulation of mucus. It is often asymptomatic (25%) and found incidentally during abdominal imaging studies or surgical explorations. Accurate diagnosis is based on clinico-pathological grounds and the treatment can differ depending on the malignant potential of the lesion.

Although appendiceal mucocele is a more common disorder than previously recognized (0.2-0.3% of appendectomies), only a few cases have been reported in the surgical literature. The prime objective of this report was to call specialist's attention about the necessity of penetrating diagnosis of appendix mucocele, a disease that is often mistaken for other abdominal pathologies. We present a case of appendiceal mucocele secondary to mucinous cystadenoma in a woman with abdominal pain, and in which the radiological study allowed the preoperative diagnosis with a high index of suspicion. The pathogenesis, clinicohistological aspects and imaging features of the lesion as well as the main therapeutic approaches suggested in the literature are commented on.

Keywords: *Appendiceal mucocele, appendicocele, mucinous cystadenoma of the appendix.*

Introduction

Appendiceal mucocele (AM) is a relatively rare entity that occurs in 0.2-0.3% of appendectomies, comprising 8% of all appendicceal tumors[1,2]. Approximately 25% of them are asymptomatic and, in most cases, founded incidentally during abdominal imaging studies or surgical explorations. There are different histologic varieties of AM: hyperplasia, cystadenoma and cystadenocarcinoma, and 10-15% of them progress to localized or diffuse peritoneal pseudomyxoma (pseudomyxoma peritonei). Definitive diagnosis is based on clinico-pathological data and the treatment may vary depending on the malignant potential of the tumor[3].

Although this entity is more frequent than it seems, only a few cases have been reported[2,3]. A case of mucocele presenting as a chronic intermittent abdominal pain is reported. Our aim was to emphasize the importance of knowing this pathology and to comment the main therapeutic approaches suggested in the literature.

Case Presentation

A previously healthy 59-year-old woman with no history of interest was admitted in our Emergency Department reporting 3 months of diffuse abdominal pain not associated with a gastrointestinal disorder, general neoplastic syndrome or other symptomatology. Physical examination revealed a large, elastic and painful mass in the right lower quadrant. Blood and urine tests results were unremarkable except for a discrete leukocytosis without left deviation. The serum concentration of carcinoembryonal antigen (CEA), Ca 19.9 and Ca 125 were not elevated. Abdominal radiography did not provide significant findings. The opaque enema was able to partially fill the mucocele (Figure 1), The mass was shown to be extra-colonic in origin, located postero-inferior to the cecum, and causing mild indentation on the caecal pole. Abdominal ultrasound showed an anterior anechoic image in the right iliac fossa and subsequent abdominal computed tomography (CT) scan reported the presence of

an elliptical 65x45x40 cm in size cystic structure dependent on the cecum, with thin enhancing wall and internal densities (Figure 2). At colonoscopy, the AM was recognized as a smooth bulbous submucosal lesion of the cecum with an impression formed by the appendiceal orifice. The valve area and terminal ileum were normal. To complete the study, gynecological exam was normal.

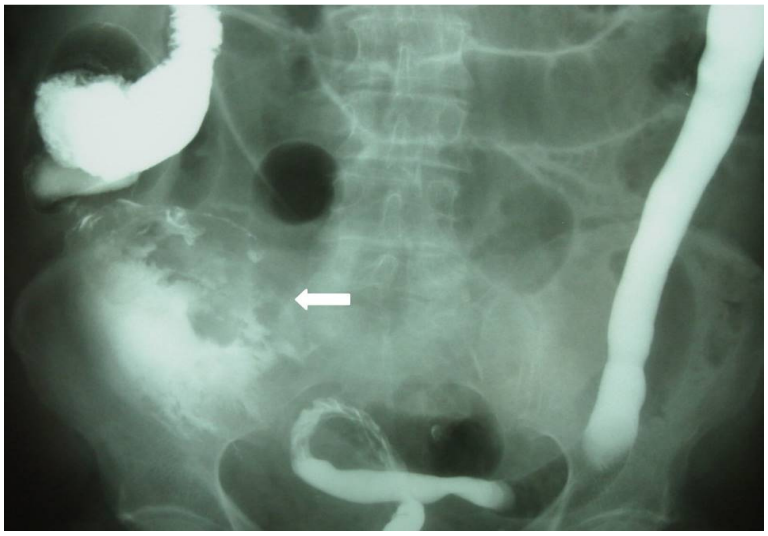

Fig 1. *Barium enema would demonstrating the presence of an endoluminal mass at the level of the cecum, right hemicolon*

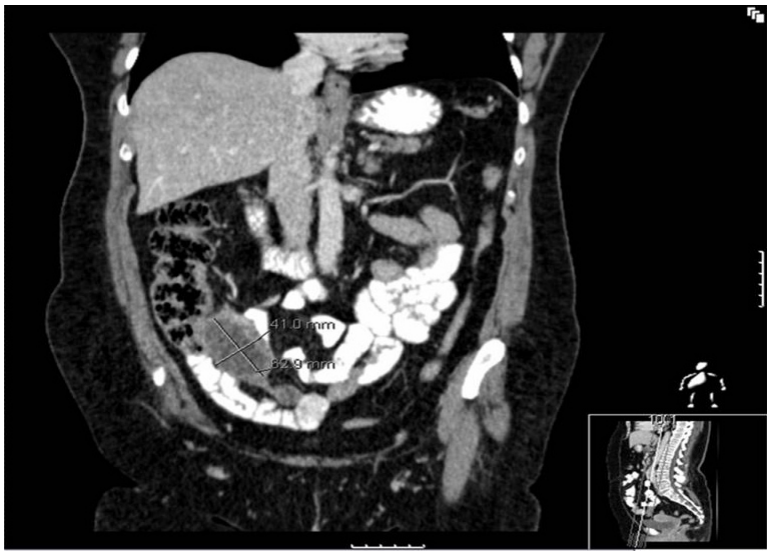

Fig 2. *Contrast-enhanced CT image shows an elliptic cystic mass in the right lower quadrant, containing low-attenuation mucin, suggestive of an appendiceal mucocele.*

Based on the clinical, analytical and radiological findings, the differential diagnosis included AM, Meckel's diverticulitis, pericolic abscess, intestinal cancer and mesenteric or hydatid cyst. With the suspicion of AM, open surgical intervention was indicated, finding a cystic mass dependent on the cecal pole with involvement of its wall (Figure 3). The rest of the colon was normal, so a right hemicolectomy was performed. The histological study showed an appendix with mucoid content and a markedly dilated light. The appendicular mucosa presented a mucosecretory epithelium with micropapillary structures and cells without atypia. In the areas of greatest dilatation, the epithelium appeared ulcerated with a giganto-cellular reaction. The muscle wall was being replaced by a fibrous tissue with secondary inflammatory reaction. Definitive diagnosis was papillary cystadenoma of the appendix with mucocele. Postoperative recovery was uneventful and currently, after 6 months of follow-up, the patient is asymptomatic and free of disease.

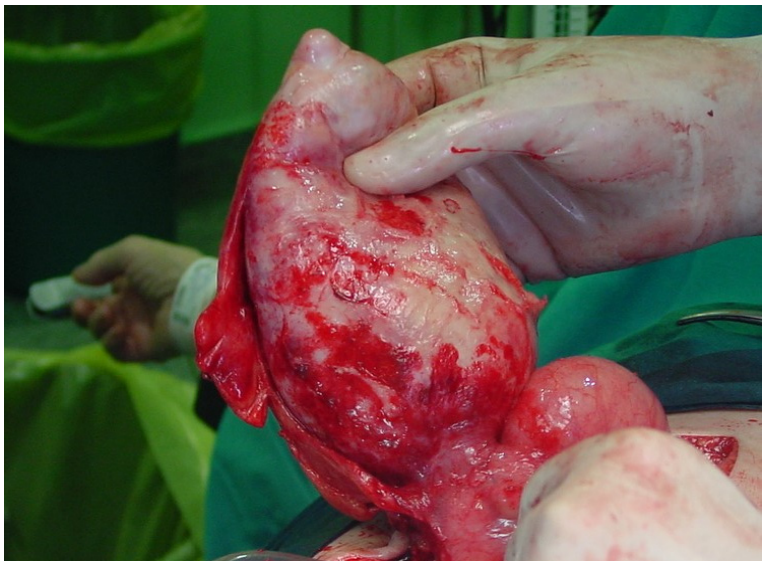

Fig 3. *Appendiceal mucocele*

Discussion

AM, defined as a dilation of the appendix due to abnormal accumulation of mucoid secretion within the lumen, was first described as "Hydrops processus vermiformis" by Carl von Rokitansky in 1842[1,4]. It shows a higher incidence in females (4: 1) and, although it can occur at any age, it usually affects adults in the 5th-6th decades[1,4]. The term mucocele is discouraged because it is merely descriptive and brings together different histological varieties with a different biological behavior, from benign forms (mucous retention cysts due to obstructive processes, focal or diffuse mucosal hyperplasia and cystadenoma, as the case in question) to malignant forms (cystadenocarcinoma and pseudomyxoma peritonei, which is characterized by the intraperitoneal accumulation of mucoid material after the tumor rupture)[1,4].

Clinical manifestations of MA include right lower abdominal pain (64%) and a palpable mass.-which are the most frequent and presents in this case- but also as alterations of the intestinal transit, acute appendicitis or incidental finding. More rarely, the clinical picture debuts as distention, intermittent colicky pain, intussusception and intestinal obstruction, gastrointestinal bleeding, abscess, urological symptoms (right ureteral obstruction) or increasing abdominal girth due to pseudomyxoma peritonei[2]. Some studies indicate that peritoneal pseudomyxoma is rarely associated with benign cystic diseases should raise a clinical suspicion of neoplasm if present[5].

In general, preoperative diagnosis is difficult, although the improvement of imaging techniques and knowledge of this type of pathology has modified this aspect. In some cases, colonoscopy can identify a yellowish mucous discharge from appendiceal orifice or the so-called "volcano sign"[2]. A plain abdominal radiograph is usually nonspecific and barium enema may show a filling defect in the cecum indicative of compression by an extrinsic globular mass.

Ultrasound typically shows a cystic mass, heterogeneous or anechoic depending on the amount of mucin, with posterior reinforcement and a well-defined wall of variable thickness, but is unable to differentiate it from acute inflammation, abscess or localized appendiceal peritonitis. On CT, AM presents as a round or tubular cystic tumor of thin and low attenuated walls in the appendiceal region, (with enhancement after the administration of contrast) similar to the rest of the intestinal loops. The presence of dystrophic calcifications in the wall, together with the absence of lymphadenopathy or infiltration of fat may be indicative of benignity, but these findings should be evaluated very carefully[2,6]. MRI features include an elongated lesion of low intensity in T1 and hyperintense in T2. In comparison, CT is more helpful than MRI in the evaluation of the region of the cecum and appendix. Due to the risk of rupture or seeding of neoplastic cells (and result in pseudomyxoma peritonei), puncture of a distended AM is generally not recommended.

The treatment must always be surgical. The technical options vary depending on the suspected histological

type or the macroscopic characteristics of the lesion[6]. Thus, benign lesions are treated and cured by performing an appendectomy while those malignant tumors or lesions that affect the cecum and those greater than 2cm in size or presenting with lymphatic invasion, serous or mesoappendix involvement must be treated with right hemicolectomy with regional lymphadenectomy. However, some authors consider that an appendectomy with resection of the cecal pole is sufficient[6,7]. In cases of emergency surgery or when it is not possible to have an intraoperative histopathological confirmation, we recommend the widest resection. In those cases in which the tumor ruptures with the consequent peritoneal dissemination of the epithelial cells and the mucous content, giving rise to the entity known as peritoneal pseudomyxoma, the indicated treatment is cytoreductive surgery proposed by Sugarbaker[5].

For this reason, careful manipulation of this type of tumor has a great importance since it has important prognostic implications. Hence, open surgery is recommended as the approach of choice, with laparoscopic surgery accepted only in expert hands, provided that the wall of the mucocele remains intact and extraction is carried out with a protective bag[2,8].

It is important to take into account the association of mucocele with other tumors, especially the gastrointestinal tract (up to 20% of cases) but also kidney, breast and ovary which makes it advisable not only the preoperative study to discard them (colonoscopy, gynecological study ...) but the periodic monitoring of these patients[8]. Survival in cases of cystadenoma reaches more than 90% at 10 years, decreasing to 25% at 5 years in cystadenocarcinomas[3,7].

References

[1] Robb H Rutledge. Primary appendiceal malignancies. In: Morris PJ, Malt RA, eds. Oxford Textbook of surgery. New York: Oxford university press; 1994. p.1117-1121.

[2] González Valverde FM, Gómez Ramos MJ, Vázquez Rojas JL. Extrinsic Ileal Occlusion Secondary to Appendiceal Mucocele of Unusual Size. Acta Chirurgica Belgica 2008; 108, (4): 454-456.

[3] Bronzino P, Abbot L, Bagnasco F. Appendiceal mucocele due to mucinous cystadenoma: case report and review of the literature. G. Chir, 2006; 27: 97-9.

[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: 1525-41.

[5] Sugarbaker PH. Pseudomyxoma peritonei: a cancer whose biology is characterized by a redistribution phenomenon. Ann Surg 1984; 219: 109-111.

[6] Soweid AM, Clarkston WK, Andrus CH. Diagnosis and management of appendiceal mucoceles. Dig Dis.1998; 16: 183-6.

[7] Misdraji J, Yantis RK, Graeme-Cook FM, Dawson. Appendiceal mucinous neoplasm: A clinicopathological analysis of 107 cases. Am J Surg Pathol.2003; 27: 1089-1103.

[8] Wolff M, Ahmed N. Epithelial neoplasm of the vermiform appendix. II: Cystadenoma, papillary adenoma and adenomatous polyps of the appendix. Cancer 1976; 37: 2511-2522.

Citation: **Francisco Miguel González Valverde, Patricia Pastor Pérez.** *Appendiceal Mucocele. Archives of Gastroenterology and Hepatology. 2018; 1(2):1-4.*