



## Original article

## Mucocele of the appendix: Presentation of 31 cases

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## A B S T R A C T

Mucocele of the appendix is a very uncommon disease estimated to be seen in 0.2%–0.3% of all appendectomies (0.28% in our series). The term “mucocele” means dilation of the appendix due to mucus, caused either by a benign process or a malignant one (cystadenoma or adenocarcinoma).

**Material and methods:** We present a series of 31 cases (17 females) treated over 18 years and with a mean age of 62 years. The most frequent clinical symptom (14 cases, 45%) was pain in the right iliac fossa of less than 72 h onset which simulates acute appendicitis.

**Results:** The histology results showed that it was benign in 21 cases. The appendix was removed in all cases; 5 by laparoscopy, with caecal resection in 8 cases (1 was a cystadenocarcinoma) and right ileocelectomy in 15 patients (9 malignant).

**Conclusion:** Follow up is recommended, not only to monitor the appendicular disease, but also due to the high incidence of synchronic or metachronic tumour processes in other areas.

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## Mucocele apendicular: presentación de 31 casos

## R E S U M E N

El mucocele apendicular es una enfermedad poco frecuente que se estima en el 0,2–0,3% de todas las apendicectomías y que en nuestra serie supone el 0,28%. El término mucocele hace referencia a la dilatación del apéndice por moco, causada tanto por un proceso benigno como maligno (cistoadenoma o adenocarcinoma).

**Material y métodos:** Presentamos una serie de 31 casos (17 mujeres) tratados en 18 años y con una edad media de 62 años. El dato clínico más frecuente fue dolor en la fosa ilíaca derecha de menos de 72 h de evolución que simulaba una apendicitis aguda, lo que ocurrió en 14 casos (45%).

## Palabras clave:

Mucocele apendicular

Tumores apéndice

Seudomixoma peritoneo

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**Resultados:** En 21 casos el estudio histológico mostró benignidad. En todos los casos se extirpó el apéndice, en 5 casos por laparoscopia, y se añadió resección cecal en 8 casos (uno era un cistoadenocarcinoma) e ileocelectomía derecha en 15 pacientes (9 eran malignos).

**Conclusiones:** Se recomienda el seguimiento, no sólo para el control de la enfermedad apendicular, sino por la alta incidencia de procesos tumorales sincrónicos o metacrónicos en otras áreas.

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## Introduction

Appendiceal mucocele is a disease with an incidence estimated at 0.2%-0.3% of all appendectomies performed and 8%-10% of all appendiceal tumours.<sup>1,2</sup>

The term appendiceal mucocele describes a dilated appendiceal lumen by a mucous secretion secondary to blockage. This increase in the lumen can cause the appendix to dilate, turning it into cystic mass that can rupture and disseminate the contents of mucin (pseudomyxoma peritonei) throughout the abdominal cavity. From a histological point of view, the concept of appendiceal mucocele includes different pathological patterns: focal or diffuse hyperplasia of appendicular mucosa, appendiceal cystadenoma, and cystadenocarcinoma.<sup>3</sup>

The most common clinical forms of presentation of this entity are: as an incidental finding during another examination, as clinical symptoms of pain or discomfort at the height of the right lower quadrant indicative of acute appendicitis, or as an abdominal mass found in the right iliac fossa. Ten percent to fifteen percent of appendiceal mucoceles evolve to a process of pseudomyxoma peritonei.<sup>3</sup>

The purpose of this retrospective study is to analyze the incidence, symptoms, treatment and survival of these rare tumours diagnosed in our hospital during the period of 1991-2008.

## Material and methods

In a period of 18 years 31 patients have been diagnosed and involved presenting a pathological diagnosis of appendiceal mucocele at the Hospital Universitario Dr. Peset (Valencia,

Spain), representing 0.28% of all appendectomies performed at our centre in this time period, a value that is within the figures found in other series. We analysed sex, age, clinical manifestations, diagnostic methods, treatment, evolution, and pathological diagnosis.

## Results

Seventeen of the diagnosed patients were women (54%). The average age of the series was 62.1 years, with intervals between 20 and 85 and a median of 67 years. The predominant symptoms were pain in the right iliac fossa at less than 72 h of evolution in 14 patients, compatible with acute appendicitis, followed by a mass in the right lower abdomen in 8 patients, longstanding pain in 6 patients and casual discovery in 3 patients.

Of the 31 patients with a pathological diagnosis of appendiceal mucocele, 16 were operated on urgently (52%) and of these, 14 diagnosed with suspected appendicitis, and another 2 with intestinal occlusion.

An abdominal ultrasound was performed on 26 patients (84%) who, like most common findings, revealed the presence of a dilated tubular structure at the height of the right iliac fossa in 14 cases, followed by a mass effect in 7 cases and intraabdominal collection in 2 cases. Tomography was used in 19 cases (61%) revealing a long tubular structure, distended with hypodense material, with or without calcification in the wall of the appendix, along with a mass effect in 8 cases. A nuclear resonance was requested in one case with suspected intraabdominal collection (3%). The colonoscopy was indicated in 10 patients (32%) which, as the most common finding, described the presence of a D-appendiceal orifice protrusion

**Table 1 – Diagnostic methodology**

| Form of presentation   | Ultrasound | CT | Colonoscopy | MRI | Barium enema |
|--|------------|----|-------------|-----|--------------|
| Acute appendicitis, n=14 (45%)   | 14         | 3  |             |     |              |
| Cecal tumour, n=8 (25%)  | 7          | 8  | 6           |     | 2            |
| Intraabdominal collection, n=3 (9%)  | 2          | 2  |             | 1   |              |
| Intestinal obstruction, n=1 (3%)   | 1          | 1  |             |     |              |
| Others: anaemia, weight loss, pain, casual finding from other cause, n=5 (16%) | 2          | 5  | 4           |     | 3            |

CT indicates computed tomography; MRI, magnetic resonance imaging.

**Table 2 – Histological surgical technique by emergency surgery**

|                                |        | Benign, No. (%) | Malignant, No. (%) |
|--------------------------------|--------|-----------------|--------------------|
| Appendectomy                   | 8 (25) |                 | 8 (100)            |
| Appendectomy + cecal resection | 7 (22) | 1 (3)           | 4 (13)             |
| Right ileal-colectomy          | 6 (19) | 9 (29)          | 4 (13)             |

at the level of the cecum (volcano-crater effect) and a barium enema in 5 patients (16%). The analytical data showed no relevant data, except leukocytosis in patients who presented acute processes (Table 1).

The treatment was surgical in all cases: an open appendectomy was performed in 3 cases and laparoscopy in 5 patients, all in benign processes and by emergency surgery. A cecal resection was added to the previous techniques in 8 cases because of diagnostic doubt or possible involvement of margins; one case was an appendicular cystadenocarcinoma in a 72-year-old woman who is free of disease and was not re-operated on. Fifteen patients underwent a right ileal-colectomy (6 of them were cystadenomas) from affection of structures adjacent to the appendix, wall rupture and presence of extraappendicular mucus or from doubts about the benignity of the process. We had no case of immediate postoperative mortality.

In 2 patients, one diagnosed with rectal neoplasm and the other diagnosed with a sigma, the finding of the mucocoele was incidental during the study of colorectal neoplasms. A left colectomy was performed in 2 patients and a colostomy in the third along with appendectomies in the same surgical time.

In 2 patients, who underwent programmed surgery and presence of free mucus throughout the abdominal cavity (peritoneal pseudomyxoma generalized) was previously identified, intraperitoneal infusion chemotherapy (mitomycin C) was added; both cases were cystadenocarcinomas (Table 2).

The anatomical pathology of the surgical specimen showed the histological benignity of the appendix in 21 cases (cystadenoma and hyperplasia) and the presence of a cystadenocarcinoma in the remaining 10. Among the benign cases, peri-appendicular mucoid material which was removed was found in 3 cases during the operation. The cytological analysis of the mucus showed material with few cells, mainly composed of macrophages and histiocytes. Of these 3 patients, one was asymptomatic, 2 died (one without identification of masses of mucus and the other due to the extension of a synchronous adenocarcinoma of the rectum). No patients were re-operated. Within the malignant cases, 5 patients died, 3 with pseudomyxoma peritonei, and the other 2 from other causes unrelated to their appendicular disease. Regarding the remaining 5, 2 of them presented intraabdominal masses with subocclusive crisis and the others are disease-free. In the malignant cases, 4 were reoperated on because of the presence of mucoid masses (pseudomyxoma) and they were removed. One of them was reoperated on in 3 cases from intestinal obstruction and the patient is currently alive.

## Discussion

The appendiceal mucocoele is an entity that may appear with a variety of clinical manifestations, sometimes even as an incidental finding in imaging tests carried out for other reasons or surgery.

Since 1842, when Rokitansky presented the mucocoele as a pathological entity,<sup>4</sup> few series have been published on this type of tumour. Of these, one of the broadest in the medical literature was published in 2003 at the Mayo Clinic with 132 patients<sup>5</sup> and, regarding Spanish authors, one published in 2007 by Ruiz-Tovar of 35 cases in a 21 year period.<sup>6</sup>

The term mucocoele refers to an enlarged appendix and by mucoid content, and it includes both benign and malignant processes, each with a different biologic behaviour. Therefore some authors, such as Higa,<sup>3</sup> discourage the use of the term mucocoele since it does not indicate whether it is benign or malignant.

In our series of 31 patients, we have found that the average age and the incidence coincide with the majority of other series<sup>5-7</sup>: this is a disease that usually occurs between the fifth and seventh decade of life.<sup>5,6,8,9</sup> However, in our series, we have a diagnosed case of focal hyperplasia in a 20-year-old woman who was operated on for suspected appendicitis.

With respect to the predominant sex, the results were very similar to other series in which no clear predominance is shown.

Regarding the symptoms presented, acute pain in the right iliac fossa, within a context of acute appendicitis, is the most common, found in nearly 45% (n=14) of cases, as is the case in other series,<sup>5-9</sup> followed by a greater distance by the existence of a mass in the right iliac fossa in 25% (n=8) and 10% (n=3) by incidental finding (2 patients during gynaecological ultrasounds and the other from urinary symptoms). The symptoms of malignant mucocoele cases were linked to weight loss, deterioration of the general condition and presence of intra-abdominal masses, whereas benign mucocoeles were more related to acute pain in the right iliac fossa. We must consider the diagnostic possibility of this disease in patients in the fifth and sixth decades of life presenting pain symptoms and a mass in the iliac fossa.

The improvement in diagnostic methods, mainly ultrasound and abdominal CT, making the preoperative diagnosis of mucocoele possible, is growing. In our series, a total of 31 cases in 21 patients were operated on with the suspicion of possible mucocoele by clinical and imaging tests. The appearance in an ultrasound of a well-circumscribed, anechoic and heterogeneous mass in the area of the appendix should raise

suspicion of this disease. A tomography of the abdomen helps to establish the diagnosis and assess the extent of the disease. The findings of a well-encapsulated cystic accumulation, sometimes with calcification of the wall in the anatomic site of the appendix, and with or without compression of adjacent structures, should suggest the disease.<sup>9,10</sup>

Without reaching the high incidence of association (nearly one third) of other malignancies that Luca Stocchi at the Mayo Clinic indicates in patients diagnosed with a mucocele, we have also observed a high percentage. Five patients had an adenocarcinoma of the colon (2 in the right colon, 1 in the transverse colon, and the remaining 2 in the sigmoid rectum, with liver metastases in these last 2). In 3 patients a mucocele was detected in the study or intervention for colon carcinoma and the rest, during follow-up. One patient who had surgery on a malignant breast tumour 5 years before, presented a double malignancy in the appendix (cystadenoma and carcinoid) and is now free of disease. We agree with other authors<sup>5,6,11</sup> that there is a high incidence of synchronous or meta-chronous malignancies in these patients, and thus, they should continue monitoring for their possible early detection. A colonoscopy<sup>12</sup> is recommended in all patients in whom there is suspicion of an appendiceal mucocele, to rule out the association of colorectal neoplasm. We must consider the differential diagnosis of benign and malignant neoplasms of the appendix, cecal region and ovaries (leiomyomas, lipomas, carcinoids and lymphomas), as well as other processes such as endometriosis, piosalpinx, ovarian cysts, mesenchymal, and so on.

An appendectomy is the therapeutic basis of treatment, because, though not proven, there may be progression of benign to malignant, or rupture of the mucocele can occur and apseudomyxoma can develop.<sup>13</sup> The surgical procedure must be related to the findings of the tumour (size, presence of local or diffuse mucus collection throughout the peritoneum, or ruptured appendix or safety margins) and its histology: a simple appendectomy is postulated in benign processes and cecal resection or right ileal-colectomy when there is involvement of adjacent intestinal segments, regional lymphadenopathy, peritoneal pseudomyxoma or malignancy.<sup>5,6,8</sup>

Although some authors contraindicate laparoscopic appendectomies and they always point to the conversion to open surgery when a mucocele is detected,<sup>14,15</sup> we think the extension of the process, the difficulty in the resection or the need for more radical surgery by the extension makes the laparotomy necessary. Looking at the indemnity of the wall of the appendix, we always extract the appendix laparoscopically, introduced into a bag, with careful handling to prevent rupture and peritoneal contamination of mucus. From 5 appendectomies performed laparoscopically, we had no intraabdominal mucus contamination. In 8 cases of appendiceal mucocele with suspected base involvement, lack of integrity on the wall or benign diagnostic doubt, we performed appendectomies with resection of the cecal pole. Regarding the size of the appendix, in all cases the thickness was greater than 2 cm, but we could not draw conclusions regarding whether malignant mucocèles presented a greater size.

The right ileal-colectomy is indicated for cases of cystadenocarcinomas when there is involvement of

adjacent structures (ileum and cecum) or the regional lymph nodes.<sup>5,6,8</sup>

We had 5 cases with diffuse peritoneal pseudomyxoma, of which 3 cases were detected in the intervention and the other 2 during follow-up. These figures correspond to 16%; similar to those reported by other authors.<sup>2,5,8</sup> The 5 cases were appendiceal cystadenocarcinomas, whereas in 3 patients with benign histological appendicular processes (one of which was synchronous with rectal adenocarcinoma) peri-appendicular mucoid material was identified. The mucoid content of pseudomyxoma may show different degrees of cellular dysplasia, which can be regarded as a form of dissemination and may significantly worsen the prognosis.

We recommend monitoring these patients not only because of the high incidence of other neoplasias, but due to the fact that there have been reports of late onset pseudomyxoma, even in a case of a benign mucocele.<sup>6,13</sup>

To conclude, there is no definite therapy for the treatment of peritoneal pseudomyxoma (peritonectomy, drainage of any collections, debulking, intraperitoneal chemotherapy, second look, etc),<sup>3,14,16,17</sup> and these cases present a high mortality rate. We, based on work by Sugarbaker,<sup>16</sup> instilled mitomycin C in 2 patients that presented intraperitoneal mucus with disappointing results.

## Conflict of interest

The authors affirm that they have no conflicts of interest.

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