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Rare and exceptional forms of acute appendicitis: Case series and literature review

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Abstract

The acute appendicitis constitutes the most frequent abdominal surgical emergency, its reputation of benign pathology is not entirely justified because one still dies of appendicitis On the one hand, its an unpredictable course which could expose the patient to serious complications; and on the other,its rare ans atypical forms, sometimes serious,masked by a simple appendicular syndrome.

For that, we report a series of 42 cases of rare appendicitis among 11000 appendectomies: 27 mucoceles,11 oxyuroses,2 tuberculosis, 1 taenia saginata and 1 cystic lymphangioma, collected over a period of 15 years from January 2004 to December 2019 jointly in the general surgery department of the military hospital of rabat and 5eme military hospital of Guelmim, we have studied the epidemiological, clinical, paraclinical, therapeutic and anatomopathological characteristics

A literature review made it possible to compare our data with those published.

After a cross-sectional comparison of our rare cases, the average age of our patients is 30ans with a male predominance in the mucocele and female in the other forms.

Clinically, a typical appendicular syndrome was dominated, ultrasound and scanner allowed to evoke the diagnosis of appendicular mucocele in 13 cases, the rest of the forms were revelealed by direct and indirect signs of appendicitis without etiological orientation.

Surgical treatment is instituted in all forms with different approaches, and medical treatment was subsequently started responding to each etiology.

The pathology examination remained the cornerstone for making the diagnosis.

Keywords: Appendicitis; Mucocel; Pinworm; Tuberculosis; Taenia Saginata; Appendicular Lymphangioma

1. Introduction

Acute appendicitis is one of the most frequent surgical abdominal emergencies. It results from an obstruction of the appendiceal lumen, leading to inflammation, vascular compromise, and, in complicated cases, perforation with risk of peritonitis. Although often considered a benign condition, it can still result in significant mortality.

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Its incidence has decreased in industrialized countries, attributed mainly to improved dietary habits and hygiene. Diagnosis remains primarily clinical, although additional examinations may be needed in cases of atypical presentation. Despite medical advances, treatment still relies on appendectomy, which remains the gold standard.

Acute appendicitis continues to pose a diagnostic challenge due to its clinical polymorphism and the lack of anatomo-clinical correlation.

The aim of this study is to analyze a large series of appendectomy cases to better characterize the epidemiological, clinical, and prognostic features of rare forms of acute appendicitis, and to assess the role of histopathological examination in their differential diagnosis.

2. Materials and Methods

This retrospective study includes 11,000 cases of appendectomies performed over a period of 15 years, from January 2004 to December 2019, focusing on 42 cases of atypical and rare appendicitis collected jointly at the General Surgery Department of the 5th Military Hospital of Guelmim and the Mohammed V Military Hospital of Rabat.

The primary objective is to identify and analyze rare histological forms of acute appendicitis to better understand their clinical presentation and impact on diagnostic and therapeutic management.

Only patients with atypical forms of acute appendicitis confirmed by histopathological examination were included. Patients with missing histological reports were excluded, even if radiological findings were suggestive.

The cases studied included: 11 cases of oxyuriasis, 27 cases of appendiceal mucocele, 2 cases of appendiceal tuberculosis, 1 case of Taenia saginata, and 1 case of cystic lymphangioma. Data were collected from medical records using a structured form with nine sections: epidemiology, medical history, consultation reason, clinical examination, paraclinical tests (biological and radiological), therapeutic management, histopathology, and postoperative course.

This study highlights the diversity of rare forms of appendicitis and underlines the importance of systematic histopathological examination for accurate diagnosis and appropriate patient management.

3. Results

This epidemiological study included 42 cases of atypical and rare appendicitis. Analysis showed that these exceptional forms can occur between the ages of 6 and 63, with a frequency peak around 30 years old. Both sexes were affected, although mucocele showed a male predominance, while the other forms were more common in women.

Regarding medical history, 65% of patients had no particular risk factors, while 35% had chronic illnesses, such as diabetes, hypertension, cardiac disorders, or von Recklinghausen disease, mainly associated with appendiceal mucocele.

Clinically, 57.5% of patients presented with a typical symptoms of appendicitis, including abdominal pain, nausea, and vomiting, while diarrhea was rare (12.5%). Four mucocele cases were completely asymptomatic and discovered incidentally.

Paraclinical investigations revealed that infectious workups were not systematically mentioned for mucoceles. Ultrasound often revealed a right iliac fossa mass or fluid collection in these specific cases. In other rare forms, ultrasound showed signs consistent with acute appendicitis. A CT scan was only performed in suspected cases or when ultrasound was inconclusive.

For surgical management, 35% of patients underwent appendectomy via McBurney incision, and 20% via laparoscopy. Right hemicolectomy was performed in 37.5% of cases, and 12.5% underwent ileocecal resection.

Postoperative outcomes were generally favorable, with no complications in 95% of cases. These results underscore the importance of tailored management for atypical forms of appendicitis and the key role of histopathology in confirming diagnosis.

4. Discussion

4.1. Appendiceal Mucocele

Appendiceal mucocele, or mucus-secreting tumor of the appendix, is a rare condition characterized by cystic dilation of the appendix due to mucus accumulation. It can affect the entire organ or just its distal portion. It accounts for 0.2–0.3% of appendectomies and is incidentally discovered in 70% of cases. It primarily affects women aged 50–60. Its benign or malignant nature remains debated [1;2].

In this study, 27 cases of mucocele were found among 3000 appendectomies (0.9%), while the literature reports an incidence of 0.1% to 0.24%. Mucocele mainly affects adults aged 50–70, with an average age of 55 according to literature [3]. In this study, the mean age was 39, ranging from 26 to 63 years.

Gender ratios vary: traditionally more common in women (4F/1M), recent studies show a male predominance. In this series, a male predominance was noted with a ratio of 1F/3M (7 women, 20 men) [4;5].

Mucocele is often asymptomatic (25% of cases) but may present as chronic right iliac fossa pain.

In this study:

- 14 patients (51.85%) had a palpable mass in the right iliac fossa.8 patients (29.63%) had a febrile acute appendicular syndrome.1 case (3.70%) was discovered during hernia repair. 4 patients (14.81%) were asymptomatic.
- Rare complications include obstruction, torsion, perforation with pseudomyxoma peritonei, or compression of adjacent organs.
- Due to its rarity, preoperative diagnosis is difficult.

Abdominopelvic ultrasound is key: it reveals a cystic mass in the right iliac fossa, hypoechoic, thin-walled, sometimes calcified, with an “onion bulb” appearance .

CT scan remains the reference test: it shows a liquid-filled appendiceal structure with calcified walls and can detect fat infiltration, lymphadenopathy, or pseudomyxoma peritonei [5].

This study compared 27 cases of appendiceal mucocele whose clinical features included palpable right iliac fossa mass (51.85%), febrile acute appendicitis (29.63%), incidental finding (3.70%), and asymptomatic (14.81%).

Preoperative diagnosis is challenging due to rarity. US shows characteristic cystic masses with "onion bulb" appearance, but CT scan (the gold standard) shows fluid-filled appendiceal structures with calcified walls. Imaging in this study was suggestive of mucocele in 48.15% of cases, appendiceal abscess in 33.33%, and was indeterminate in 18.52%.

Four histologic types exist: retention cyst (18%, benign), mucosal hyperplasia (20%, benign), mucinous cystadenoma (52%, benign), and mucinous cystadenocarcinoma (18%, malignant).

Management is histology-dependent - benign cases only require appendectomy (85-100% 5-year survival), while malignant cases need right hemicolectomy (25-38% 5-year survival). Ruptured cases with peritoneal dissemination may require extensive operation, peritoneal lavage, and chemotherapy, including hyperthermic intraperitoneal chemotherapy in pseudomyxoma peritonei. [6;7].

Figure 1 Appendectomy specimen case of appendiceal mucocele

4.2. Appendiceal Oxyuriasis

Enterobius vermicularis is the cause of oxyuriasis, a common worldwide intestinal parasitic disease that can lead to appendicitis, although this is not generally accepted. Incidence in specimens of appendicectomy is 0.35% to 12.5%. [8].

Mainly affects children and young adults under the age of 15 years with mild female predominance.

Pain in the right iliac fossa in all (100%), sometimes with nausea, vomiting, and diarrhea. Fever is noted in ~18% of the patients. McBurney's sign is always positive, with moderate leukocytosis (11,500-18,000/mm^3), raised CRP, and patchy eosinophilia. Alvarado scores are usually less than in classical acute appendicitis. [9-11].

Imaging shows typical findings of appendicitis but no specific signs of oxyuriasis. Laparoscopic appendectomy is appropriate (72% of cases), but there is a risk of peritoneal dissemination of pinworms. Histopathology confirms the presence of pinworms in the appendiceal lumen (90% of cases).

Insecticide treatment with two doses of albendazole is needed post-operatively. Household treatment with Vermox® or Fluvermal® for recurrence is indicated. Proper hygiene practice is necessary for prevention.

4.3. Appendiceal Tuberculosis

Appendiceal tuberculosis is a rare condition that makes up only 0.26-2.3% of appendectomies and occurs through three mechanisms of infection: intraluminal spread (most common), hematogenous dissemination, and contiguous spread from the adjacent organs. [12].

Three anatomo-clinical forms - intermittent pain, vomiting, diarrhea (chronic); phlegmonous appendicitis (acute); incidental histological discovery (latent). Two morphologic forms - hypertrophic (pseudo-tumoral) and ulcerative (with abscess/perforation).

35 years old with female predominance, though in this series, younger patients (mean 18 years, range 17-19) with even gender ratio (1:1) were found.

Pain in the right iliac fossa accompanied by nausea and fever is the primary symptom. Peritoneal irritation in 50% of the patients and McBurney's sign are invariably positive. Leukocytosis (15,300-16,000/mm^3) and elevated CRP are found in laboratory reports.

Laparoscopic appendicectomy is done in addition to WHO-guideline anti-tuberculosis treatment for nine months (two months intensive phase on four drugs, followed by seven months consolidation on two drugs). Diagnosis is established by histopathology by epithelioid giant-cell granulomas with caseous necrosis. Prognosis is excellent with complete recovery and no complications reported. [13–15].

Figure 2 Surgical specimen, case of appendicular tuberculosis

4.4. Appendiceal Taenia saginata

Appendiceal involvement of Taenia saginata is very uncommon, and few cases have been reported in the last 50 years. The literature reveals a mean patient age of 28.7 years (16-51) with female preponderance. [16–19].

The patients typically present with right iliac fossa (RIF) pain, nausea, vomiting, and fever, with abdominal guarding and positive McBurney's sign - features shared across reported cases and the patient in this study.

Imaging demonstrates evidence of acute appendicitis, sometimes with appendicoliths or peri-appendiceal collections. Management is straightforward appendectomy (via McBurney incision or laparotomy) followed by single dose praziquantel antiparasitic therapy.

Although most have uncomplicated recoveries, the patient in this report presented with a postop right iliac fossa abscess 12 days later, which was drained under CT guidance. An enterocutaneous fistula formed through which Taenia filaments were noted to protrude and were extracted. The patient was given niclosamide therapy and recovered uneventfully.

Prevention of Taenia saginata infection relies on proper cooking of beef : whole cuts to ≥63°C (with rest for 3 minutes), ground beef to ≥71°C. [19].

Examination confirms Taenia saginata infection, with the rare report of co-infection with other parasites like Enterobius vermicularis. Parasitological examination definitively identifies the species.

4.5. Cystic Lymphangioma of the Appendiceal Mesentery

Cystic lymphangiomas (CLs) are benign malformations that are derived from embryologic anomalies of the lymphatic system. While they are most commonly located in the head and neck, they occur in the abdomen rarely (2-8% of cases) and appendiceal involvement is rare.

CLs may occur in any age, but 40% are diagnosed within the first year of life and 80% by age six. Few reported cases of appendiceal occurrences are in young adults, without a uniform gender distribution. [20–22].

Patients present with repeated abdominal pain, sometimes with nausea and mild fever. Right iliac fossa tenderness, a positive McBurney's sign, and palpable abdominal mass on physical examination are evident. Non-specific presentation is rare to raise preoperative suspicion, and presenting ones are with ill-defined abdominal pain, appendicular syndrome, or rarely intestinal obstruction, peritonitis, or intra-abdominal hemorrhage.

Laboratory results usually exhibit leukocytosis. Ultrasound demonstrates hypoechoic cystic masses with thin walls and swelling of the appendix. CT yields precise anatomical details, whereas MRI is useful for characterizing cyst fluid content. [23,24].

Complete surgical removal by laparotomy or laparoscopy is the preferred treatment. McBurney incision simple appendectomy may suffice in mild cases. It is established on histopathology showing multilocular cystic masses lined by endothelium. The prognosis is excellent with no recurrence on follow-up of one year.

5. Conclusion

Appendectomy is the most frequently performed emergency surgical procedure. This retrospective study of 11,000 appendectomies performed over 15 years identified 42 rare cases of atypical appendiceal conditions (mucoceles, oxyuriasis, tuberculosis, one Taenia saginata case, and one cystic lymphangioma).

Although these patients clinically presented with typical signs of acute appendicitis, only histopathological examination enabled accurate diagnosis. The study emphasizes the crucial importance of systematic histological analysis to detect these rare forms, even when clinical presentation appears conventional, ensuring precise diagnosis and appropriate management.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study

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