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Giant mucocele of the appendix in pregnancy: A case report and review of literature

Lateef O. Idris^{a,*}, Olaniyi O. Olaofe^b, Olaniyi M. Adejumbi^a, Adedayo O. Kolawole^a, Ademola K. Jimoh^a^a Department of Surgery, Ladoke Akintola University of Technology Teaching Hospital, Osogbo, Nigeria^b Department of Pathology, Ladoke Akintola University of Technology Teaching Hospital, Osogbo, Nigeria

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ABSTRACT

INTRODUCTION: Mucocele of the appendix is an obstructive dilatation of the appendix by intraluminal accumulation of mucoid material. Mucoceles may develop by one of four processes: Retention cysts, Mucosal hyperplasia, Cystadenomas, and Cystadenocarcinomas. The clinical presentation of a mucocele is nonspecific and often an incidental finding at operation for acute appendicitis. If mucocele is allowed to rupture either spontaneously or during surgery, the escape of mucin and epithelial cells into the peritoneal cavity – Pseudomyxoma Peritonei – may occur.

PRESENTATION OF CASE: We present the case of a 35 years old pregnant woman, who was admitted to the accident and emergency department with history and ultrasound findings suggestive of acute appendicitis. The ultrasound scan also confirmed a viable fetus at about 23 week's gestational age. She had an open appendectomy. The intra-operative findings were an enlarged, tense, cystic retro-caecal appendix which was about 14 cm × 5 cm × 3 cm in dimensions. Histopathological examination confirmed mucocele of the appendix. She had normal vaginal delivery at term and postoperative follow up for 1 year was uneventful.

DISCUSSION: Mucocele of the appendix is an uncommon disorder with nonspecific presentation ranging from asymptomatic disease diagnosed incidentally to clinical features of appendicitis. The treatment is surgery.

CONCLUSION: Appendiceal mucocele is difficult to diagnose preoperatively, and a giant size of the simple mucocele may be connected to hormonal influence of pregnancy. In the index case, as in majority of cases, the definitive diagnosis is made at surgery.

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1. Introduction

Appendiceal mucocele (AM) is a rare disease of the appendix that occurs as a result of an obstructive dilatation of the appendix due to intraluminal accumulation of mucoid material. The incidence ranges from 0.2% to 0.7% of all appendectomy specimens, with a higher incidence in females and in people above the age of 40 years [1–4]. Appendiceal mucocele may result from one of four processes: Retention cysts, Mucosal hyperplasia, Cystadenomas, and Cystadenocarcinomas. The clinical presentation of a mucocele is nonspecific, and often an incidental finding at operation for acute appendicitis, as it was in this case [1,3,4]. Rupture of AM with escape of epithelial cells into the peritoneal cavity or the malignant change of the epithelium with resultant growth through the deeper layer

onto the peritoneal surface where it becomes implanted, can give rise to pseudomyxoma peritonei (PMP), which has a high mortality rate [1,4]. Other complications of AM include torsion, inspissations with subsequent calcification and formation of stones [4]. Because of the risk of PMP, when a mucocele is visualized at the time of laparoscopic examination, conversion to laparotomy is recommended, so as to ensure that a benign process is not converted to a malignant one through mucocele rupture [3]. The principles of surgery include resection of the appendix, wide resection of the mesoappendix to include all the periappendiceal lymph nodes and complete evacuation of the intraperitoneal mucoid material for cytologic examination.

2. Case report

A 35-year-old G4P3⁺⁰ (3 alive) woman was admitted to the accident and emergency at twenty-three weeks gestational age with 3 day history of gradual onset colicky peri-umbilical abdominal pain. The pain became severe, about 4 h prior to presentation. She had two episodes of vomiting preceded by nausea and anorexia. There was no history of fever or change in bowel habit. On examination,

* Corresponding author at: General Surgery Unit, Ladoke Akintola University of Technology Teaching Hospital, Osogbo, Nigeria. Tel.: +234 8035959693.

E-mail addresses: ldrisol@yahoo.com (L.O. Idris), rindeolaofe@yahoo.com (O.O. Olaofe), adejumbi43@gmail.com (O.M. Adejumbi), cullerwarley@yahoo.co.uk (A.O. Kolawole), yuabim@yahoo.com (A.K. Jimoh).



Fig. 1. Excised appendix (along with the base of the caecum) measuring 14 × 5 × 3 cm.

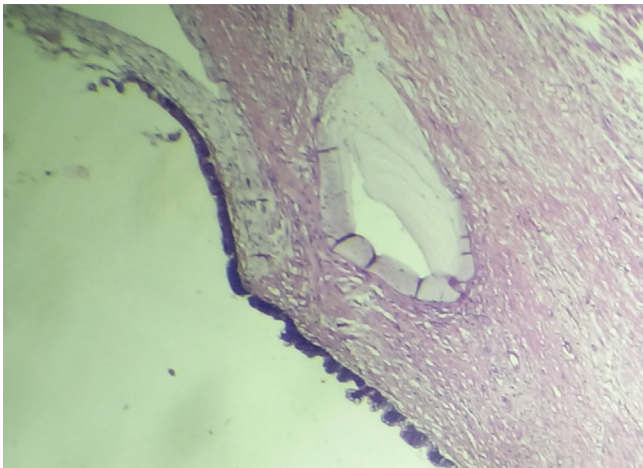


Fig. 2. Section (H&E x40) showing mucinous columnar epithelium. There is presence of mucin within the wall and reactive inflammatory response.

she was in painful distress, afebrile (axillary temperature of 37.1 °C) and not pale. Abdominal examination revealed about 22 week's size gravid uterus with moderate and rebound tenderness in the right iliac fossa. She had an abdominal ultrasound scan which was suggestive of acute appendicitis. The ultrasound also confirmed a viable gestation. Full blood count and serum electrolyte values were within normal limits. Based on the preoperative diagnosis of acute appendicitis in pregnancy, she had emergency open appendectomy under spinal anaesthesia and through Lanz incision. The intra-operative finding was an enlarged, tense, cystic retrocaecal appendix. The base of the appendix was bulging into the caecum (Fig. 1).

The wall was grossly inflamed but there was no demonstrable perforation, no enlarged peri-appendiceal lymph nodes or any other pathology. Diagnosis of mucocoele of the appendix was suspected. She then had excision of the appendix, mesoappendix and the base of the caecum followed by simple closure of the caecum. She was discharged home on post operative day 4. Histopathological examination confirmed simple mucocoele of the appendix (Figs. 2 and 3).

She had normal spontaneous vaginal delivery at term. A one year postoperative follow up at the surgical outpatient and postnatal clinic was uneventful.

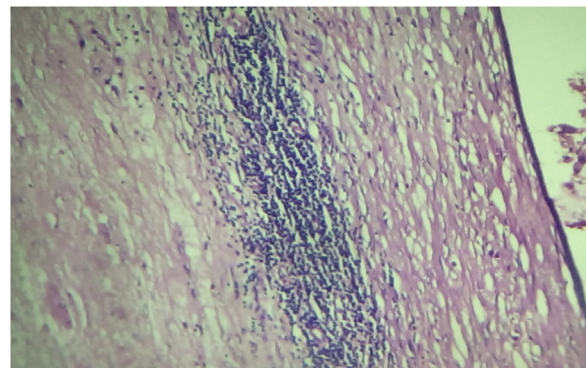


Fig. 3. Section (H&E x40) showing a thin flattened mucosa with mucous columnar epithelium. Lymphoid aggregates are also seen in the wall.

3. Discussion

Appendiceal mucocoele (AM) refers to a dilatation of the appendiceal lumen caused by the accumulation of mucous material irrespective of the primary pathology that has led to this [4,5]. AM was recognized as pathological entity by Rokitsky in 1842 and was formally defined by Feren in 1876 [5,6]. AM is divided into 4 pathological subgroups based on the epithelial characteristics [2–5]:

- i) Retention cyst or simple mucocoele: it develops following obstruction of the lumen of the appendix usually by faecolith. It has normal or flattened epithelium, moderate luminal dilatation of up to 2 cm and it constitutes about 20% of all appendiceal mucocoeles. The case presented falls under this group. However, it was much larger than expected for this pathological type. This may suggest the possibility of hormonal influence due to the pregnant state of the patient.
- ii) AM with hyperplastic epithelium and moderate luminal dilatation: this constitutes about 20% of AM.
- iii) Cystadenoma: this is characterized by tubular adenomatous epithelium with varying degree of epithelial atypia. It produces large amounts of mucin with prominent luminal dilatation of up to 6 cm. It is the most common form, constituting about 50% of cases and with associated 20% risk of perforation.
- iv) Cystadenocarcinoma: this type is characterized by invasion of the glandular stroma and may be associated with the implantation of epithelial cells in the peritoneum. It sometimes resembles mucinous carcinoma of the colon. It constitutes about 11–20% of all cases with 6% risk of spontaneous rupture [5,7,8].

Cystadenoma and Cystadenocarcinoma are neoplastic appendiceal mucocoeles and they constitute about 35% of all primary neoplasms of the appendix [5,9]. These lesions may occur de novo or from preexisting simple mucocoeles. In these conditions, complete excision of the appendix is usually curative especially if the appendix is excised intact and histopathological examination confirms the absence of malignant cells at the margin (negative margin). However, in cases of rupture, deposition of mucous material in the peritoneal cavity occurs, resulting in tumour dissemination. The gelatinous mucin substance later forms a semisolid mass in which the malignant glandular cells are found: this is known as pseudomyxoma peritonei (PMP) [5,10]. PMP can also result from the growth of epithelial cells (after assuming malignant potential) through the deeper layers onto the peritoneal surface where they become implanted [4,5]. Several investigators, now believe that cystadenoma undergoes malignant transformation, evolution of which may be similar to that of the adenoma–adenocarcinoma sequence of the colon [5,8]. Therefore,

the disease can undergo insidious progression that may be unnoticed for years. Metastasis from Cystadenocarcinoma is rare. It tends to remain in the peritoneal cavity, although few cases of retroperitoneal and pleural implantations have been reported [5,6,11,12].

Clinical features range from asymptomatic disease to various manifestations. These presentations include symptoms and signs of acute appendicitis, long history of painless or painful right iliac fossa (RIF) mass, abdominal distension with discomfort (if PMP has occurred), features of intestinal obstruction from mucocele associated intussusceptions, incidental findings of calcified RIF mass on plain abdominal radiograph and a well circumscribed mass displacing the caecum medially on barium enema. AM can be an incidental finding at laparotomy for other conditions [1,4,5]. One third of patients with AM have associated localized tumours in the gastrointestinal tract (especially colorectal tumours) and tumors of the ovary, breast, and kidney.

Preoperative diagnosis may be possible if there are pathognomonic signs like onion-skin-like circles on ultrasonography or if abdominal computerized tomography reveals a round, low-density, thin-walled, encapsulated mass communicating with the caecum and calcifications which may be present in about 50% of cases [5,13–16].

Colonoscopy may reveal a characteristic mass with a central crater from which mucoid material exudes. This is known as “volcano sign” [5,14–17].

Treatment is surgical, but laparoscopic approach is not advised because of the risk of rupture [2,3,18]. The presence of a mucocele of the appendix does not necessitate performance of a right hemicolectomy. The principles of surgery include resection of the appendix, wide resection of the mesoappendix to include all the appendiceal lymph nodes and peritoneal collections, cytologic examination of all intraperitoneal mucus, and careful inspection of the base of the appendix to rule out any extension into the caecum. Right hemicolectomy, or preferably caecectomy, is reserved for patients with positive margins at the base of the appendix or positive periappendiceal lymph nodes. Recently, a more aggressive approach to ruptured appendiceal neoplasms has been advocated which involves primary resection of the tumour with all gross implantations [2,3,18,19]. This approach includes a thorough but minimally aggressive approach at initial laparotomy, consideration for re-exploration if indicated and hyperthermic intraperitoneal chemotherapy (HIPEC) [3,5].

Postoperatively, patients with simple or benign neoplastic mucocoeles have shown an excellent prognosis with 5-year survival rates of 91–100%, even in cases with extension of mucus into the extra-appendiceal spaces. In malignant mucocoeles, however, the 5-year survival rate is markedly reduced (25%) due to complications of pseudomyxoma peritonei [2,20].

4. Conclusion

Appendiceal mucocele is difficult to diagnose preoperatively and a giant size of the simple mucocele may be connected to hormonal influence of pregnancy. In the index case as in majority of cases, the definitive diagnosis is made at surgery.

Conflict of interest

We (the authors) hereby write to disclose that we have no financial or personal relationships with other people or organizations that could inappropriately influence (bias) this work.

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We (the authors) hereby declare that we have no competing interests.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Idris O.L. performed the operation, conceived the design, and participated in the literature review; Olaofe O.O. did the pathologic examination; Adejumbi M.O. and Kolawole O.A. participated in literature search and revision of this write up; and Jimoh K.A. assisted the operation and participated in the follow up of the patient.

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