



Case Report:

Mucocele of Appendix Secondary to Cystadenoma a Diagnostic Challenge

Prasad K. Shetty, Ramesh M, Saraswathi Ramesh,

Department of Pathology, Obstetric and Gynecology and Surgery,
AV Hospital, Bangalore, India

Address For Correspondence:

Dr. Prasad. K. Shetty,

Surgical Pathologist,

AV Hospital, Patellamma temple Road

Basvangudi, Bangalore, India

E-mail: dr.pkshetty@gmail.com

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Abstract:

Mucocele of appendix is uncommon cystic lesion characterized by distension of the appendiceal lumen with mucus. Most of them are caused by mucinous cystadenomas and rarely cystadenocarcinomas. Clinical presentation is varied with more than half being asymptomatic. We report such a case where initial clinical findings and investigations suggested an ovarian cyst, and the diagnosis was only made at the time of surgery. In women presenting with a right iliac fossa mass and clinical features not indicative of gynaecological pathology, an appendiceal origin should be considered in the differential diagnosis.

Key Words: Appendix; Mucocele; Ovarian cyst

Introduction:

Mucocele of the appendix secondary to mucinous cystadenoma is a rare clinical finding. They can present as a pelvic mass and thus pose a diagnostic challenge.

Currently, the assessment of right iliac masses relies heavily on ultrasonography as the primary diagnostic tool. This however may not always identify the origin of such a mass. In spite of preoperative investigations, the diagnosis may still remain elusive and may only be made at the time of surgery.

Case Report:

A 53 yr old woman came with history of right-sided abdominal pain, dull and intermittent in nature since 2 months. Her past history is insignificant. On physical examination, her abdomen was flat, soft, with normoactive bowel sounds. Tenderness was noted in the right upper and lower abdomen without rebound pain or muscle guarding. On pelvic examination, hard, mobile mass approximately 6x4 cms was palpated in the right iliac fossa. Her serum CA 125 was 6.4 IU/ml (Normal <35 IU/ml). An ultrasound scan showed a right sided mixed echogenic pelvic mass with an echogenic rim, possibly ovarian in origin, measuring 61 × 43 × 51 mm.(Fig. 1)



Figure 1: Ultrasound scan showing right sided pelvic cystic mass.

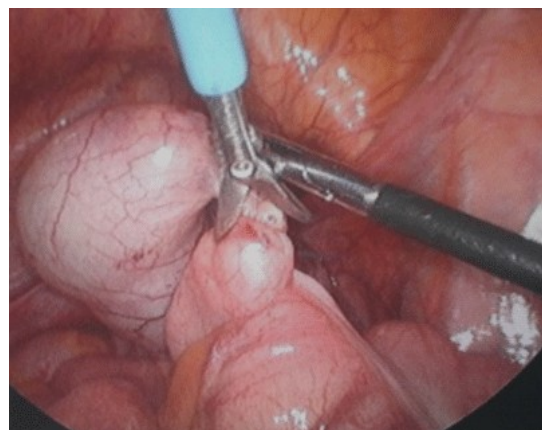


Figure 2: Intraoperative photograph of cystic mass arising from appendix.

A laparoscopy was planned with the diagnosis of pelvic mass. A soft cystic mass with a diameter of approximately 80 x 80 mm with a smooth surface originating from the appendix in the pelvic region was identified and a routine appendicectomy was performed.(Fig. 2) Abdominal irrigation was performed

and abdominal viscera were evaluated as normal. Grossly, appendix was globularly enlarged measuring 6x4cms, outer surface was smooth, on cut opening appendix was cystically dilated filled with large amount of mucin, wall was 6mm thick.(Fig 3)



Figure 3: Gross photograph of cystically dilated appendix filled with abundant mucin

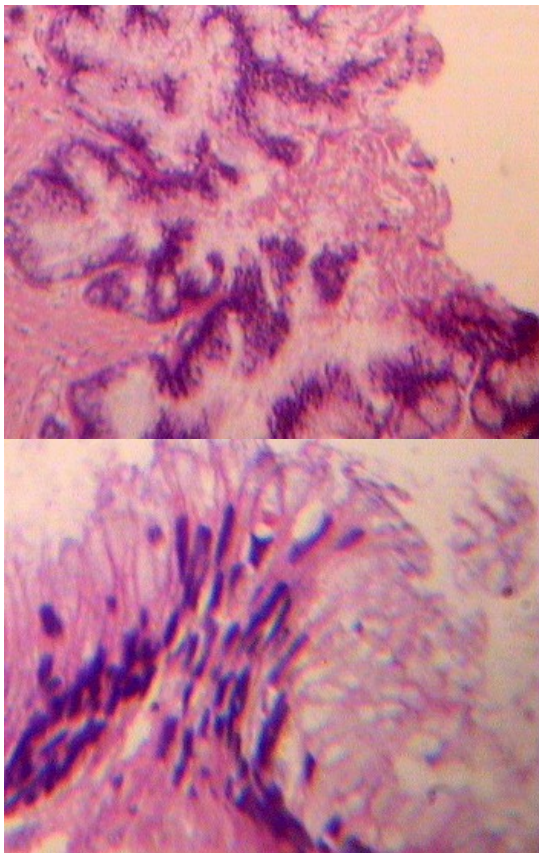


Figure 4,5: 10x and 40x microphotograph showing cystadenoma appendix lined by atypical mucinous lining epithelium

On Microscopy appendix showed atypical lining mucinous epithelium predominantly in a papillary configuration, lining cells were stratified columnar cells with basal, elongated, hyperchromatic nuclei, with abundant apical mucin, muscularis propria is thinned out with hyalinization.(Fig. 4,5)

Discussion:

Mucocele of the Appendix make up about 0.2%–0.3% of appendicectomies. Appendiceal mucoceles show a female pre-

dominance of 4:1. The average age at the time of diagnosis is 54 years for benign mucoceles and 64 years for malignant disease.^[1] The most common presentation of symptomatic appendiceal mucocele patients is acute or chronic right lower quadrant abdominal pain, as occurred with our patient.^[2,3] An intra-abdominal mass is palpated by the examining physician in half of cases.^[4] Nausea and vomiting, as well as altered bowel habits are often reported.^[5,6] An association between appendiceal mucocele and synchronous colon neoplasms has been previously noted. The most common synchronous neoplasms occur in the large bowel (19.5%-21%).^[7] A correct pre-operative diagnosis of appendiceal mucocele is difficult due to the nonspecific symptoms.^[8] Computed tomography (CT), ultrasonography (US), and colonoscopy have all been used to describe these tumors. Unfortunately none of these is entirely conclusive.^[1,8] In our case ultrasonography could not provide a preoperative diagnosis.

Surgical excision of mucocele of appendix can be either by laparotomy or Laparoscopy. Laparoscopic surgery provides the advantages of good exposure and evaluation of entire abdominal cavity, as well as more rapid recovery with avoidance of a large incision and a better cosmetic outcome. However careful handling of the specimen is recommended as spillage of the contents can lead to pseudomyxoma peritonei.^[9,10]

Histopathologic classification of appendiceal mucoceles is dependent on the characteristics of their lining epithelium; these include retention cysts (18%), mucoceles with mucosal hyperplasia (20%), mucinous cystadenomas (32%), and mucinous cystadenocarcinomas (10%). Classification is important; because the course of the disease and prognosis are related to these subtypes.¹⁰ Simple mucoceles (retention cysts) are characterized by degenerative epithelial changes and may result from appendiceal obstruction and distension. There is no evidence of hyperplasia or neoplasia of the mucosa. Hyperplastic mucoceles are sessile or pedunculated lesions that represent hyperplastic polyps of the colon and are not known to have any malignant potential. Mucinous cystadenomas also have been referred to as low-grade appendiceal mucinous neoplasms. They typically are circumferential cystic lesions composed of mucin rich epithelium. While mucinous cystadenomas can be considered the equivalent of adenomatous colon polyps. The 5-year survival rate for simple or benign neoplastic mucocele ranges from 91% to 100%.^[5-7]

In conclusion mucocele of appendix is very rare. It is very difficult to diagnose preoperatively in women as it mimics adnexal mass. The mucocele of the appendix must be considered in patients with advanced age; especially in female, with atypical ultrasonographic appearance; or a right adnexal mass.

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