MUCOCELE APPENDIX - CASE REPORT AND LITERATURE REVIEW

ISHTIAQ AHMED¹, MUHAMMAD SALMAN², SUNDAS ISHTIAQ³

INTRODUCTION

Rokitansky first described mucocele of the appendix in 1842.¹ It is a rare pathology characterized by the appendix lumen dilatation with mucus accumulation. It is more common in the 5th and 6th decade of life, affecting predominantly female and found in 0.2% to 0.3% of the appendectomy specimens.²⁻⁶ Etiopathogenesis can be inflammatory or neoplastic. Mucocele Appendix may be due to cystadenomas (63%), cystadenocarcinomas (11%), mucosal hyperplasia or inflammatory process (25%) and rarely due to lumen obstruction by carcinoid tumors or endometriosis.⁷ In addition to these causes, other tumor of appendix or caecum may present as mucocele. The main complication of mucocele is pseudomyxoma peritonei.⁸⁻⁹ In our case, the mucocele typically presents as right lower quadrant abdominal pain and vomiting which favor the clinical diagnosis of acute appendicitis. The symptoms did not assist in making the pre-operative diagnosis of mucocele appendix. The objective of this review is to analyze literature as to mucocele, especially regarding diagnosis and treatment, besides discussing follow-up and prognosis of the individuals who have this disease.

CASE HISTORY

A Thirty Six year old woman was referred by GP with one day history of right sided abdominal pain, vomiting and anorexia. There was no associated urinary, bowel or gynecological symptoms. She had same complaints in past off and on for last about six years which improve after medical treatment by her GP. She was febrile, mildly dehydrated, having tenderness and guarding over right iliac fossa. No mass or abdominal viscera are palpable. Her blood complete picture shows raised TLC (11200/cmm) with neutrophilia, and US scan shows a suspected mixed echogenic tubular mass of about 1.5 cm thickness suggestive of acute appendicitis. She underwent surgery and per-operatively the appendix was normal, but grossly dilated (up to 3cm) with broad base (Figure - 1). Due to lack of frozen section facilities, partial caecal resection along with excision of fat and lymph nodes of mesoappendix was done after extending the incision. She had smooth post operative recovery and her histopathology report shows simple benign mucocele appendix. The patient had been repeatedly visiting her local GP’s and Tertiary care hospitals where she was constantly being misdiagnosed and sent back on conservative management.

DISCUSSION

The preoperative diagnosis of mucocele is usually difficult due to nonspecific clinical presentation of the mucocele in appendix.³⁰ Mostly the patients are asymptomatic (23-50%) and usually it is diagnosed incidentally during Surgery.³¹ Occasionally, it may be diagnosed during endoscopic procedures, ultrasound or radiological examination of gut.³² ³⁵ Right lower abdominal pain is the commonest clinical presentation (50%) which persists for months without any palpable mass.³² A change in bowel habit or rectal bleeding is rarely reported by patients.³³ Simple or benign mucocele rarely perforate due to insidious progression as compared to malignant mucocele which have sudden onset like acute appendicitis which sometime present as an organ perforation.³⁴⁻³⁸ Radiology, Ultrasonography or endoscopy may facilitate preoperative detection especially when this pathology is suspected by clinician. An ultrasound finding varies depending on the contents from purely hypo echoic cystic lesion or
An anechoic fluid having fine internal echoes or complex hyper echoic masses. The “onion skin sign” is considered to be specific for mucocele of the appendix. 

CT scan of the abdomen, sometime shows a well-encapsulated cystic mass communicating directly with the caecum. Rarely, curvilinear or mural punctate calcification without any surrounding inflammatory reaction causing extrinsic pressure on the caecal wall is seen. These findings confirm the mucocele diagnosis and also differentiate it from appendicular abscess. 

Perforated mucocele appendix may lead to the pseudomyxoma peritonei. In this situation, peritoneum usually act as a defense against the dissemination of mucous, so it is recommended that the initial surgery should be as minimal as possible, in order to keep the peritoneum intact. 

Cytoreductive surgery and intraperitoneal chemotherapy is recommended in cases with limited dissemination in peritoneal cavity. Literature review of 123 patients with pseudomyxoma peritonei from a single institution showed that the five year survival is reported 50% to 96%, in selected cases who has completed cytoreduction and there are no distant metastases. 

CONCLUSION

Although mucocoele appendix is a rare pathology, but its diagnosis should be kept in mind during appendicectomy because proper management can lead to a better outcome.

REFERENCES


