MUCINOUS CYSTIC TUMOUR OF APPENDIX INVOLVING CAECUM - A RARE PRESENTATION

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ABSTRACT:

Mucocele of the appendix is an infrequent event , comprising 0.3-0.7% of appendiceal pathology and 8% appendiceal tumours. It is characterized by a loculated or diffuse distension of appendix with a mucus filled lumen. The main differential diagnosis would include mesenteric cyst, duplication cyst, right ovarian cyst and hydrosalpinx. This case is about a 52year old female with vague, chronic pain abdomen, more on right side for 2 years ; ultrasound was suggestive of a right adnexal mass and explorative laprotomy showed a mucinous cystic tumour of appendix.

KEY WORDS: *Appendix, mucinous tumour, irritable bowel syndrome, cysadenocarcinoma, laparotomy, right hemicolectomy*

INTRODUCTION:

Mucinous cystic tumour of appendix is a very rare entity , with overdistension of the appendix with mucous secondary to luminal obstruction by fecolith or foreign body , carcinoid or endometriosis . Mean age of occurrence is around 55 years of age with four times female preponderance. It is mostly associated with colonic adenocarcinoma or a mucin secreting tumour of ovary. Clinically, 25% of patients are asymptomatic and the rest present with acute or chronic right lower quadrant abdominal pain. Pre-op diagnosis of underlying malignancies in a mucocele is important for the management. However, its identification is difficult on imaging studies.

CASE REPORT:

A 52 year old multiparous female presented to our outpatient department with complaints of occasional right sided pain abdomen for 2 years, which has been aggravated for the past 2 months. There was no history of fever, vomiting, loss of appetite and weight loss. She had no bladder disturbances and there were no features suggesting pelvic inflammatory disease. She was a known case of irritable bowel syndrome for past 9 years on treatment. No other major illness or surgeries in the past except for transabdominal tubecotomy.

Examination of her systems were within normal limits and vaginal examination revealed a fullness in the right fornix which was cystic to firm in consistency, and with normal sized uterus. Basic investigations were done which was within normal limits. Ulrasound-Pelvis was suggestive of a linear right adnexal mass with a possible diagnosis of right sided hydrosalpinx. Patient was taken up for diagnostic laparotomy and to proceed for further management.

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Intraoperatively, uterus, bilateral tubes and ovaries were within normal limits. Appendix was enlarged in size. Hence appendicectomy was performed and the specimen sent for histopathology confirmed a borderline mucinous tumour of the appendix and with base involving the caecum (Fig. 1).

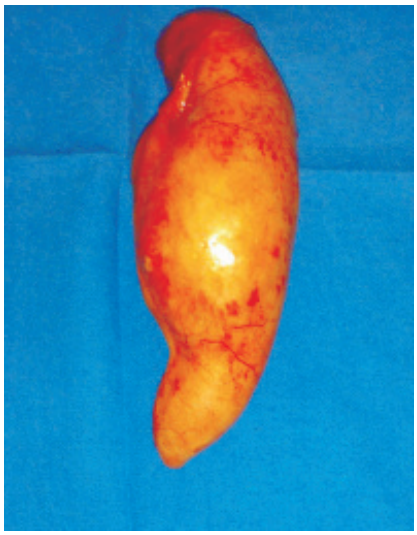

Fig. 1 : Appendiceal Mucocele

There was no evidence of ascites and no signs suggestive of liver metastasis. In consultation with surgeons, a right hemicolectomy, with terminal ileum 15cms upto right one third of transverse colon resection with ileotransversal colostomy anastomosis (Fig. 2). Intra-operative and Post-operative period was uneventful.

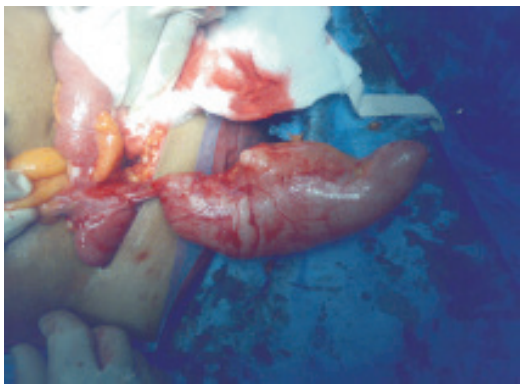

Fig. 2 : Mucocele of the Appendix

DISCUSSION:

The prevalence of appendiceal mucocele is about 0.4% among appendicectomies[1]. It is considered as mucinous distension of the appendiceal lumen regardless of the underlying pathology.

Four causal pathological conditions include [2]:-

(1) Retention cyst

(2) Mucosal hyperplasia

(3) Cystadenoma (of unknown malignant potential)

(4) Cystadenocarcinoma

Clinical manifestations include chronic or acute onset pain abdomen as in our case. It can be associated with palpable abdominal mass, gastrointestinal bleeding, weight loss, nausea, vomiting, changes in bowel habits and anemia. One third are associated with gastrointestinal tumours.

Diagnosis is very difficult. About 60% of cases are diagnosed in the intra-operative period. Pre-operative diagnoses with ultrasound and Computed topography is of utmost importance [3]. Ultrasound (USG) could show an elongated echogenic mass with concentric and echogenic layers within cystic mass (ONION SKIN) appearance [4].

Surgical resection is the method of choice for appendiceal mucocoele. Explorative laparotomy with or without right hemicolectomy is performed as in this case. Laproscopic resection might lead to peritoneal dissemination. The 5 year survival rate according to various other case reports suggests that it was 68% (Right hemicolectomy) versus 20% (appendicectomy alone)[5] .

Inflammatory bowel disease and irritable bowel syndrome (as in our case) is associated with elevated risk of certain gastrointestinal cancers (mucocele of appendix)[6]. Primary adenocarcinoma of appendix is uncommon, it is often an incidental finding and the pre-op diagnosis is very important Characteristic cystic mass in right lower quadrant and with the appearance of "ONION SKIN SIGN" (by USG) clinches the diagnosis (Fig. 3).

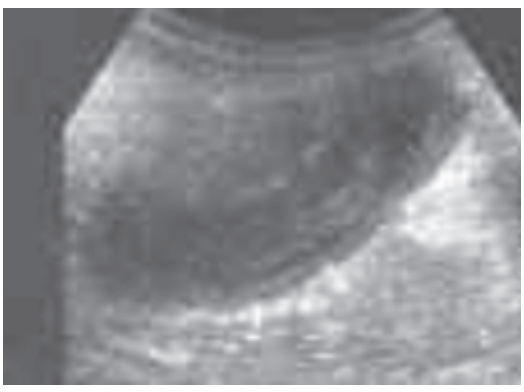

Fig. 3 : Onion Skin Sign in Ultrasound

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