Volume : 4 | Issue : 7 | July 2015 • ISSN No 2277 - 8179

Research Paper

Medical Science

KEYWORDS : Mucocele of appendix, papillary adenocarcinoma, retroperitoneal mucus collection, atypical presentation

An Atypical Presentation of Papillary Adenocarcinoma of Caecum',

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ABSTRACT

Mucocele of appendix is seen rarely in appendectomies. Out of these, very few are associated with colorectal tumours with pseudomyxoma formation. These tumours are mostly mucinous cystadenomas or cystadenocarcinomas. We present a very rare case where a 54 year old patient presented with psoas abscess following appendectomy for mucocele of appendix; incision and drainage was done; after a few months, he again came with mucous discharge from a sinus in the right lumbar region and later found to have papillary adenocarcinoma of caecum with retroperitoneal collection of mucoid material

INTRODUCTION:

The term 'Mucocele of the appendix' was coined by Karl Freiherr von Rokitansky1 in 1842. It is found in about 0.072 to 0.633% of all appendicectomy specimens. Mucocele is a broad term describing a collection of mucoid material inside the appendix. The aetiology is diverse4: most commonly, retention mucocele, villous hypertrophy causing excess mucous production, mucinous cystadenoma or mucinous cystadenocarcinoma of the appendix or caecum, or even endometriosis of the appendix.

We present a case where a well-differentiated papillary adenocarcinoma of caecum was found in a patient operated for mucocele of appendix 30 months earlier.

CASE DESCRIPTION:

A 54 year old male patient first presented to our emergency department in September 2012 with pain in the right iliac fossa. He was diagnosed with appendicitis and appendectomy was done. Operative finding was mucocele of appendix but histopathological report was not corroborative. Post-operative recovery of the patient was uneventful and he was discharged on the 4th post-operative day and was later lost to follow-up till the next visit.

In December 2014, the patient again presented with history of pain in the right lumbar region. Clinically there was fullness in the right flank. Ultrasonography showed a psoas abscess, which was drained through extra-peritoneal approach using a gridiron skin incision. Contents and wall scrapings of the cavity were sent for histopathological examination to rule out tuberculosis. The HPE report revealed inflammatory tissue consistent with diagnosis of pyogenic abscess. The track of the abcess healed over a month.

The patient came back in April 2015 with complaint of discharge of pus-like material from a sinus that formed posterior to the earlier grid-iron incision. The patient also had fullness to the right of the umbilicus.

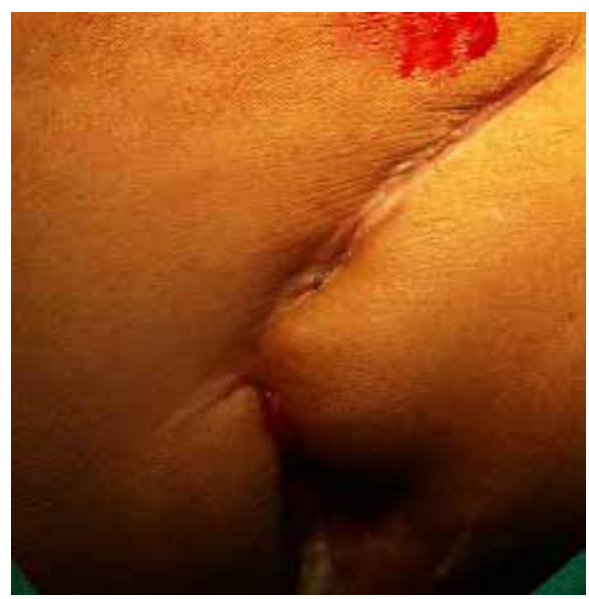

FIGURE 1. DISCHARGE FROM SINUS THAT FORMED POSTERIOR TO THE EARLIER GRID IRON INCISION (Viewed from the right with patient in supine position)

Ultrasonography showed a 9x5cm sized hypoechoic collection in the right lumbar region anterior to the psoas muscle. Diagnosis of recurrent psoas abscess was made. Under spinal anaesthesia, sinus was reopened through gridiron incision. Mucoid material was found in the retroperitoneal cavity. Suspecting pseudomyxoma peritonei, mucoid material was evacuated as much as possible and wall scrapings were obtained for sending for histopathological examination. Corrugated rubber drains were placed in the cavity through the sinus track and the incision was closed.

The patient was then subjected to colonoscopy which showed polypoid non-ulcerative growth. Biopsy was taken and sent for HPE, which showed dysplastic epithelium with adenomucinosis.

Volume : 4 | Issue : 7 | July 2015 • ISSN No 2277 - 8179

However, Serum Carcino-Embryonic Antigen (CEA) level was normal.

Patient was posted for prophylactic right hemicolectomy. On laparotomy, an annular irregular growth was found in the caecum and distal ileum, with collection of mucoid material in retroperitoneal region.

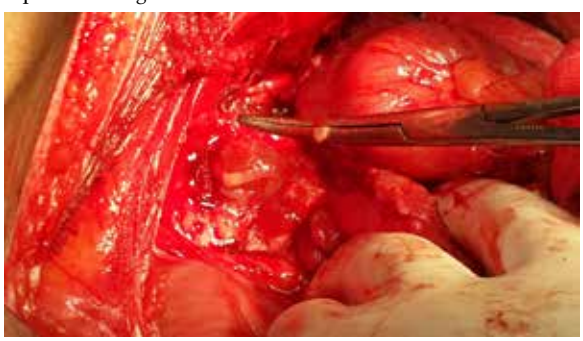

FIGURE 2. MUCOID MATERIAL IN THE RIGHT ILIAC FOSSA FOUND AT LAPAROTOMY

Right hemicolectomy was done and the mucoid collection was evacuated. Pathological examination revealed papillary adenocarcinoma of the caecum and distal ileum with TNM staging of T3NxMo.

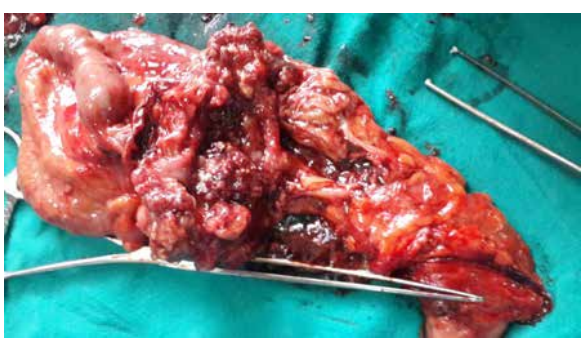

FIGURE 3. RIGHT HEMICOLECTOMY SPECIMEN WITH MASS IN CAECUM AND DISTAL ILEUM

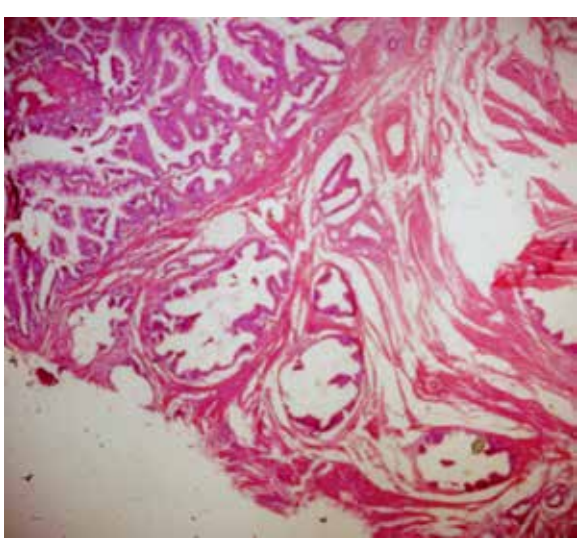

FIGURE 4. MICROSCOPIC VIEW OF THE PAPILLARY ADENOCARCINOMA

DISCUSSION:

The term mucocele of appendix refers to local or diffuse dilatation of the lumen of appendix due to abnormal accumulation of mucus. As mentioned above, the etiology varies widely, from retention of mucus to neoplasms. It is usually seen in females above the age of 55. Most of these tumors are clinically silent, however they may present with acute or chronic right lower quadrant pain, bowel obstruction, bleeding and intussusception[5].

Mucocele may be associated with presence of acellular mucin in the peri-appendiceal region or with dissemination of mucus-producing cells into the peritoneal cavity causing pseudomyxoma peritonei in some cases.

Misdraji et al[6] classified mucinous appendicular tumors into two groups: low-grade appendicular mucinous neoplasms (LAMN) characterized by villous and/or flat- mucinous epithelial proliferation and a low degree of atypia, and mucinous adenocarcinoma defined by tumoral invasion of the bowel wall, complex epithelial proliferation and high-grade nuclear atypia.

Many cases of mucocele have been reported to be associated with concomitant neoplasms, most commonly adenocarcinoma of the appendix[7], caecum, ascending[8] or descending9 colon, and also of the ovary causing an adnexal mass.

But association of mucocele with papillary adenocarcinoma is very rare and has been seldom reported in the literature. Furthermore, in our patient, post-appendectomy, a psoas abscess had formed, probably due to post-surgery infection, which after incision and drainage, remained as a potential space for collection of mucin. At the last visit, we first suspected it to be a recurrence of the psoas abscess, as colonoscopy could identify only a non-ulcerative polypoid mass but no evidence of any other growth in the large intestine. Serum CEA level was also found to be normal. However, the retrieved wall scrapings specimen was found to have dysplastic changes and hence right hemicolectomy was done. The final report showed well-differentiated papillary adenocarcinoma.

The significance of CEA levels in diagnosis is doubtful, though it is very useful in prognostication of colorectal tumours. Hence, even in cases with borderline CEA values, there must be a high index of suspicion and other investigations must be carried out to make sure that a malignancy can be safely ruled out.

CONCLUSION:

Our case emphasises that mucoceles are complex in nature and can have varied etiologies and presentations. There can be associated tumours of the intestine, ovary, etc., ranging from benign to malignant. At present, there are no guidelines for estimating the malignant potential or prognosis for these tumours. There is no consensus regarding the best line of treatment either. Hence, each case should be investigated thoroughly, considering every option possible in order to arrive at the correct diagnosis and to identify the treatment regimen that would be most suitable for the patient.

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