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Mucinous Cystadenoma Leading to Giant Mucocele of Appendix

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Abstract:

Mucocele of appendix is a rare entity characterized by cystic dilation of the appendix due to intraluminal accumulation of mucoid substance resulting from chronic obstruction of appendix orifice due to any inflammatory or neoplastic cause. Pre-operative diagnosis is obligatory to differentiate it from cyst adenocarcinoma. We report a case of giant mucocele which presented as asymptomatic abdominal lump, with brief review of literature regarding its clinical, diagnostic and characteristics.

Key words: Adenocarcinoma, Mucocele, Cysts, Appendix.

Introduction

Mucocele of appendix is a rare entity characterized by cystic dilation of the appendix due to intraluminal accumulation of mucoid substance. Incidence wise, it constitutes only 0.2%-0.4% of total appendectomies. It is reported to be more common in females (4:1) and elderly (> 50 years) [1-3]. Mucocele results from chronic obstruction of appendix orifice due to any inflammatory or neoplastic cause. On the basis of etiopathogenesis, mucocele's are divided into four histological types, retention cysts, mucosal hyperplasia, mucinous cystadenomas and cyst adenocarcinomas. Mucocele of size less than 2 cm are usually associated with retention cyst and mucosal hyperplasia. Larger mucocele are more likely to be neoplastic [4]. Mucinous cystadenoma is responsible for maximum number of mucocele (63%). Histopathologically, it exhibits hyperplastic adenomatous mucosa. Mucinous cystadenoma is a benign condition and adequately treated by a simple appendectomy. Pre-operative diagnosis is obligatory to differentiate it from cyst adenocarcinoma because more radical surgery is required for latter and its rupture can lead to a grievous outcome in from of pseudomyxoma peritonei.

We report a case of giant mucocele presented as asymptomatic abdominal lump, with brief review of literature regarding its clinical, diagnostic and characteristics.

Case Report

A 54 year male presented with asymptomatic abdominal lump in right lower quadrant. Patient consulted a physician 20 days back for coryza and

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during that visit, he was noted to have a palpable abdominal lump. There was no history of abdominal pain or any other complaints related to lump. On examination, the lump was confined to right iliac fossa. Lump was firm, painless, non-mobile, with smooth surface and well defined margins. Ultrasound revealed cystic mass arising from the caecum base with non-visualization of appendix. Computed tomography of abdomen showed a large lobulated, well defined hypodense lesion showing no contrast enhancement in close proximity of caecal wall.

Patient was then posted for exploratory laparotomy which demonstrated a cystic mass of size 14x6 cm, involving almost whole of the appendix except for 1 cm of the base. It was partially adherent to cecum and was later separated by blunt dissection [Fig.1]. As mucocele was not invading the base and meso-appendix and there was no evidence of lymphadenopathy, simple appendectomy was done. Cut section of specimen showed gelatinous material [Fig.2].

Histological finding was of cystic structure lined by mucin secreting epithelium with focal papillary in-folding suggestive of mucinous cystadenoma with mild atypia. Patient recovered well without any complication.

Discussion

Mucocele of the appendix was recognized by Rokitansky in 1842 [1]. An appendicular mucocele leads to progressive enlargement of the appendix. The various histopathological lesions and percentage of overall mucocele are mucosal hyperplasia and retention cyst (25%), mucinous cystadenoma (63%-84%), and mucinous cyst adenocarcinoma (11-20%) [3-8].

About 25% of patients remain asymptomatic due to slow-growing distension of the

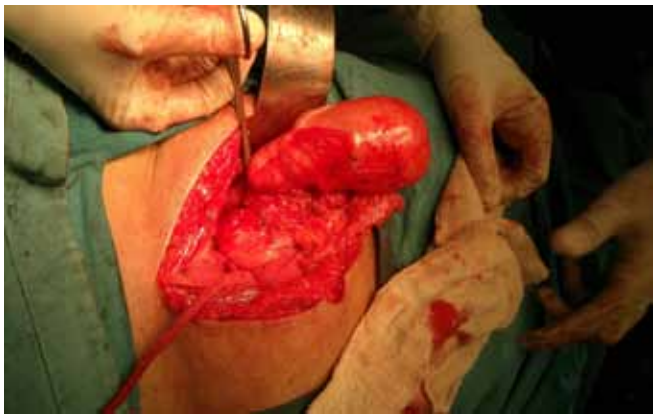

Fig.1: Mucocele of appendix.

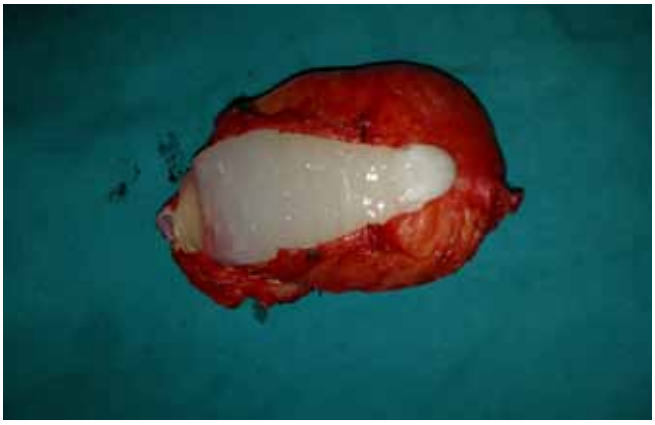

Fig.2: Cut section of mucocele of appendix.

appendix and present incidentally on physical or radiological examination, as in our case [2]. Clinical manifestations include right lower abdominal pain, palpable abdominal mass, gastrointestinal bleeding and features of intestinal obstruction due to intussusception or volvulus [2]. Abdominal pain is the most common symptom followed by abdominal lump, which is present in 50% of patients. 23% to 50% of mucoceles are detected incidentally during laparotomy for undiagnosed acute abdomen [3].

Diagnosis of a mucocele can be established by ultrasound or abdominal CT scan. Ultrasonography usually shows a cystic, encapsulated lesion, in relation to the caecum with variable internal echogenicity

and calcification of the cyst wall. CT scan finding of a mucocele include a well encapsulated, thin walled cystic mass and nodularity suggests possibility of cyst adenocarcinoma. Calcification is seen in 50% of cases [8,9]. Colonoscopy can demonstrate 'volcano sign' i.e. soft erythematous mass, with a central crater due to the protrusion of appendicular orifice [7]. The carcinoembryonic antigen (CEA) level at preoperative may suggest malignity in the appendix or in the colon [1]. The differential diagnosis should include carcinoid tumor, adenocarcinoma, lymphoma, peri-appendiceal abscess, and ovarian tumors.

For mucinous cystadenoma, appendectomy is sufficient if the lesion does not involve the appendicular base. Occasionally, the mass will rupture prior to or at the time of removal, but this rupture is typically contained to the right lower quadrant and is considered localized pseudomyxoma peritonei. If the mass is benign, appendectomy and removal of any residual mucin is curative. Laparoscopic appendectomy is not currently recommended due to the possibility of malignancy and spillage of mucin-secreting cells throughout the abdomen [2]. Gonzalez-Moreno *et al*. recently reviewed 501 patients diagnosed with appendicular epithelial neoplasm and justified right hemi-colectomy when there is necessity to clear the primary tumor, ileocolic lymph node involvement demonstrated by histopathological examination and a non-mucinous neoplasm identified by histopathological examination [9].

Conclusion

Patients harboring giant mucocele can be asymptomatic. Superlative management of mucocele is achieved through accurate preoperative diagnosis and subsequent careful resection.

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