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MUCINOUS CYST OF THE APPENDIX

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Abstract : A 64 yrs male patient who came for general health check up was found to have a right iliac fossa mass of size 17cm by 8cm on CECT. D-Lap showed a mucinous cyst arising from tip and body of appendix, with the caecum and base of appendix free from tumour. Open appendicectomy was done. The histopathology report came as mucinous cyst without evidence of dysplasia. Patient is still followed up with USG and colonoscopy.

Keyword :mucinous cystadenoma, appendix, appendiceal tumours, mucinous cystadenocarcinoma

INTRODUCTION:

The term mucocele simply refers to a cystic mass filled with mucin, as in a dilated appendix. Simple mucoceles secondary to inflammatory obstruction do occur with or without atrophic epithelial changes. These are benign entities without pathologic significance. However, a more descriptive classification system based on the changes to the underlying appendix epithelium has been proposed to try and define biologic behavior. These histopathologic lesions and percentage of overall mucoceles are then defined as MUCOSAL HYPERPLASIA (25%), MUCINOUS CYSTADE-NOMA (63%) and MUCINOUS CYSTADENOCARCINOMA (12%).1 Appendiceal mucoceles is a rare but well recognized entity that can mimic several common clinical syndromes or present as incidental surgical or radiological finding. It has a 0.2% - 0.4% prevalence among appendicectomies.

CASE REPORT:

A 64 yrs old male patient presented to general surgical OPD for a routine general health check up, and was found to have a heteroechoeic cystic mass of size 13.9cm across 4.9cm with cyst wall in the right iliac fossa by USG Abdomen. He was asymptomatic and denied of weight loss, fever, flushing symptoms or change in bladder and bowel habits. On examination abdomen was soft with a doubtful

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University University Journal of Surgery and Surgical Specialities vague mass in right iliac fossa. CECT showed well encapsulated mass of size 17cm by 8cm in right iliac fossa without adjacent structure involvement. Appendix and ileo-caecal junction could not be visualized. Colonoscopy was found to be normal. A provisional diagnosis of lymphoma was made and was planned for diagnostic laparoscopy, which showed a mucinous cystic lesion involving the tip and body of the appendix. Caecum and base of the appendix was found to be normal, hence open appendicectomy done. The cyst along with appendix was removed in to without spillage of the content. There was no other intraperitoneal cystic lesion. Histopathologic analysis of the mass was mucinous cyst of appendix. Section showed fibrocollagenous tissue of cyst wall lined by tall columnar epithelium with apical mucinous cytoplasm. There was no evidence of malignancy in the material studied. Repeat USG and colonoscopy after three months was normal and the patient is still being followed up.

DISCUSSION:

The differential diagnosis of a cystic right lower quadrant mass is extensive and includes primary adenocarcinoma, carcinoid tumour, mucinous cystadenoma, mucinous cystadenocarcinoma, lymphoid hyperplasia, lymphoma, peri-appendiceal abscess, and in females, ovarian tumours. When symptoms do not suggest an inflammatory, hemorrhagic, or acute obstructive process, ancillary studies may help to refine the diagnosis. The most common presenting symptom associated with mucinous cystadenoma has been abdominal pain; however, one-fourth of patients are asymptomatic and are found incidentally.2 Complications like bleeding, intussusception, and local invasion into surrounding structures are described with associated symptomatology.3-5 By ultrasound, cystic masses with varying internal echogenicity and a layered wall with calcification may be seen. Computed tomography findings show well-encapsulated cystic masses with low attenuation, usually without associated appendiceal inflammation. Varying thickness of the mass wall did not seem to correlate with malignancy, although contrast-enhancing nodules may suggest cystadenocarcinoma.

Computed tomography does help in evaluating other pathologic processes. Colonoscopy is usually nondiagnostic, as mucosal biopsies will often be normal. Suggestion of extrinsic compression or mass protrusion of the appendiceal orifice can be helpful.

Histologically, the classification scheme for appendiceal mucoceles outlines a spectrum of epithelial changes that are analogous to changes in the colon. Mucosal hyperplasia and mucinous cystadenomas are akin to colonic hyperplastic polyps and adenomatous polyps.6 Concomitant cases of mucinous cystadenomas and colon cancer have been described with an incidence of up to 20%.1,7 The progression to malignancy has not been proven for mucinous cystadenomas, but it is suggested. The intraoperative distinction between benign mucinous cystadenoma and mucinous cystadenocarcinoma remains difficult without the clinical picture of wall invasion, local spread, or widespread peritoneal implants (pseudomyxoma peritonei). This can be made more difficult in the face of acute inflammation. Histologic analysis is not always straightforward and relies on evidence of stromal invasion and marked nuclear atypia of the appendix epithelium to diagnose mucinous cvstadenocarcinoma.

The morbidity/mortality associated with both cystadenoma and cystadenocarcionoma stems from rupture and intraperitoneal spread of mucin-producing epithelium. No reports of lymphatic or hematogenous spread are found. It is apparent that both mucinous cystadenocarcinomas and cystadenomas have the capacity to produce pseudomyxoma peritonei, although the clinical burden is likely dependent on the degree of dysplasia.

Regardless of the ultimate histology, extreme care must be taken in tissue handling. Masses in the appendix body without local invasion or ceacal involvement can be treated with simple appendicectomy and mesoappendix excision. Most seem to advocate completion right hemicolectomy if final histology proves cystadenocarcinoma. Any mass involving the caecum or adjacent organs should be resected by a right hemicolectomy. Intraoperative exploration of the entire gastrointestinal tract and ovaries in females is warranted. All gross peritoneal implants should undergo a biopsy and be removed with grading of the degree of epithelial atypia for prognostic purposes.8

In our case, we chose initial diagnostic laparoscopy to assist with operative planning. It allowed assessment of the mass and evaluation of the remainder of the abdominal cavity. Ultimately, a mass freely involving the appendix body could be treated by a standard open approach as for appendicectomy. The ability to safely handle tissues and adequately assess the caecum for the lesion extent is a clinical decision left up to the operating surgeon.

Cystic lesions of the appendix especially mucinous cystadenoma and mucinous cystadenocarcinoma are difficult to diagnose despite extensive preoperative evaluation. Although the surgical treatment is straightforward, proper management of the incidentally found lesion requires understanding of the potential complications of widespread peritoneal disease. Thorough intraoperative gastrointestinal, ovarian, and peritoneal examination is required. Follow-up colonoscopy and pelvic examination is also warranted for the high association with other colon and ovarian malignancies.

CONCLUSION:

Appendiceal mucocele presents as a challenge to the surgeon who does not appreciate the effect of pathological diagnosis on the operative procedure. Appendicectomy alone should be performed only on mucinous lesions that are determined to be non-neoplastic after biopsy. If appendicectomy is performed, precautions should be taken to minimize the risk of seeding the peritoneal cavity with tumourous mucin during manipulation. Right hemicolectomy is recommended for cases in which the pathologic diagnosis has not been established and for any cases suspicious of adenocarcinoma or complicated by concomitant adenocarcinomatous disease or pseudomyxoma peritonei. Preoperative evaluation with radiologic and endoscopic methods is helpful but not always diagnostic.

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