



STUDY ON INCIDENCE AND MANAGEMENT OF APPENDICULAR MALIGNANCY IN PATIENTS UNDERGOING APPENDICECTOMY IN A TERTIARY CARE HOSPITAL

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ABSTRACT

AIMS: To study the incidence and management of appendicular malignancy in patients undergoing appendicectomy in a tertiary care hospital

METHODS: A retrospective study including 1,275 patients who were admitted in JSS hospital, under surgery department, between September 2016 to September 2021. Patients were ≥ 16 years, any gender who presented with pain abdomen, abdominal distension, fever, vomiting, or mass per abdomen, diagnosed as appendicitis or appendicular mass. Patients underwent laparoscopic/open appendicectomy or right hemicolectomy, based on intraoperative findings. The specimen was sent for histopathological examination and followed up for further management if required.

RESULTS: Among 1275 patients who underwent appendicectomy either open or laparoscopy, following histopathological examination of all the appendicectomy specimens, appendicular malignancy was reported in 7 patients (0.53). All 7 patients had complaints of pain in the right iliac fossa. On examination, 71.4% had tenderness in the right iliac fossa. Most common preoperative provisional diagnosis was acute appendicitis (57.1%). 3 patients underwent laparoscopic appendicectomy, 1 open appendicectomy, and based on intraoperative findings, 2 underwent right hemicolectomy, 1 resection and anastomosis with ileostomy as metastasis was present. Histopathologically, 2 patients were diagnosed with adenocarcinoma appendix, 2 with low grade appendiceal mucinous neoplasm, 2 with metastatic mucinous adenocarcinoma and 1 patient with neuroendocrine tumour of appendix

CONCLUSION: Appendiceal tumors are uncommon, majority are comprised of epithelial neoplasms. Present with abdominal pain, manifest as acute appendicitis, or incidentally discovered in appendectomy specimens during histopathological evaluation. Imaging findings are suggestive of acute appendicitis mostly. CT is used for staging of appendiceal tumors and subsequent follow up. In majority, appendicectomy will be sufficient, and some require right hemicolectomy. Cytoreductive surgery and intraperitoneal chemotherapy have improved the prognosis of patients with peritoneal disease.

KEYWORDS

Appendicular malignancy, laparoscopic appendicectomy, Cytoreductive surgery

INTRODUCTION

Primary and secondary neoplasms of the appendix are rare tumors found in approximately 1% of appendectomy specimens (Connor et al., 1998). Common appendiceal tumors are epithelial neoplasms and neuroendocrine tumors (NETs). Other tumors are rarely encountered and include lymphoma, metastases, neuroectodermal and nerve sheath tumors, mesenchymal tumors. There are no established risk factors for the development of appendix cancer. Malignancy of appendix most often present with acute appendicitis and diagnosed incidentally on histopathological assessment of the surgical specimen (Kelly, 2015). Appendix cancers may also be asymptomatic and be found incidentally on colonoscopy, surgery, or on cross-sectional imaging for other indications. Patients usually present with pain abdomen, abdominal distension, fever, vomiting episodes or anorexia. May present with abdominal distension and pain secondary to peritoneal dissemination. Best tools for investigation is ultrasonography, or CT abdomen for further evaluation. Preoperatively the most common differential diagnosis made is acute appendicitis or appendicular mass, for which appendicectomy is planned, and based on intraoperative findings other surgical options considered with histopathological examination of the specimen.

METHODS

This was a retrospective study which includes 1,275 patients who were admitted in JSS hospital, under surgery department during, September 1st 2016 to September 1st 2021. These patients presented with pain or mass abdomen, abdominal distension, fever, vomiting, and were admitted, evaluated, diagnosis of appendicitis or appendicular mass was made. Patient underwent either laparoscopic/ open appendicectomy or right hemicolectomy, based on intraoperative findings. The specimen was sent for histopathological examination and followed up for further management if required.

RESULTS:

Amongst 1275 patients who underwent appendicectomy either open or laparoscopically, histopathological examination of all the appendicectomy specimens reported appendicular malignancy in 7 (0.54%) patients, with remaining 99.46% who underwent appendicectomy were diagnosed with benign conditions of the appendix

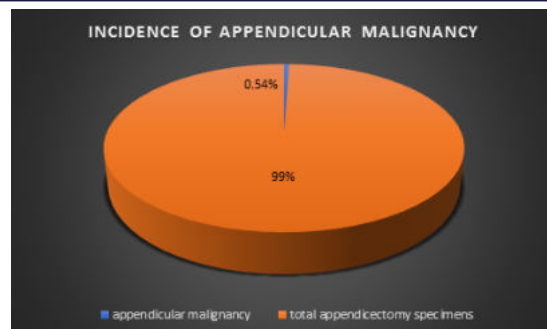


Figure 1: Incidence of appendicular malignancy

Out of the 7 patients diagnosed with appendicular malignancy, 4 were females (57.1%) and 3 were males (42.8%), hence showing a female preponderance in the incidence of appendicular malignancy. Amongst the 7 patients who were diagnosed with appendicular malignancy, all 7 patients had complaints of pain abdomen, mainly in the right iliac fossa, with 71.4% having loss of appetite and anorexia, 57.1% having complaints of vomiting episodes and loss of weight. 28% presented with abdominal distension along with pain abdomen, and 1 patient presented with complaints of fever and constipation.

Amongst the 7 patients, diagnosed with appendicular malignancy, their preoperative examination revealed presence of anaemia, ascites, tenderness in the right iliac fossa, and mass in the right iliac fossa. 71.4% of the patients diagnosed with appendicular malignancy patients were examined to have tenderness in the right iliac fossa, 57.1% with mass in the right iliac fossa region, 42.8% with anaemia. Amongst the 7 patients detected with appendicular malignancy, provisional diagnosis was acute appendicitis in 4 patients (57.1%), appendicular mass in 3 patients (42.8%), and suspected carcinoma caecum or suspected ileocaecal tuberculosis in one patient.

Of the 7 patients who underwent surgery, 3 patients underwent laparoscopic appendicectomy, 1 patient underwent open appendicectomy, 2 patients who were planned for appendicectomy underwent right hemicolectomy in view of intraoperative findings, and 1 patient underwent resection and anastomosis with ileostomy in view

of intraoperative findings of metastasis, diffusely involving the omentum and peritoneum.

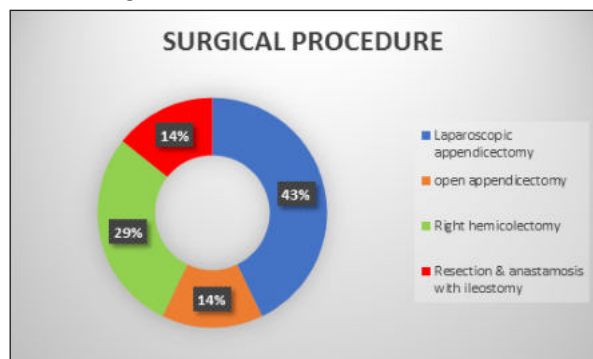


Figure 2: Surgical procedure

Out of 1275 patients who underwent surgery, 7 patients were detected with appendicular malignancy, based on the histopathological examination of the specimens obtained from surgical excision. Of these 7 patients, 2 patients were diagnosed with adenocarcinoma appendix, 2 with low grade appendiceal mucinous neoplasm, and 2 patients with metastatic mucinous adenocarcinoma. One patient was detected with NET of appendix.

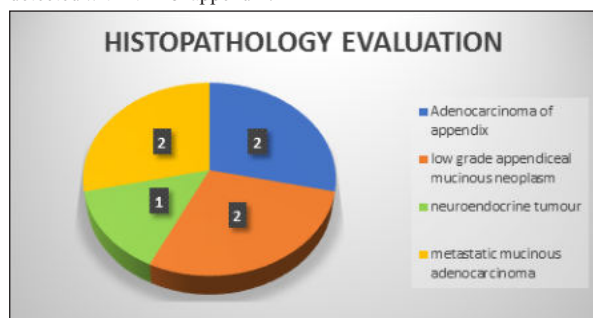


Figure 3: Histopathological evaluation of specimen

DISCUSSION:

Primary and secondary neoplasms of the appendix are rare tumors found in approximately 1% of appendectomy specimens^(Connor et al., 1998). Appendiceal adenocarcinomas occur frequently in middle-aged or older adults, with a median age at presentation in the 4th and 5th decades being most common malignant neoplasm of the appendix, while malignant NETs are seen in younger patients in the 2nd decade. The most frequent initial manifestation of appendiceal tumors is acute appendicitis, seen in 30%–50% of patients and more commonly in NETs than in epithelial neoplasms^(Connor et al., 1998; Deshmukh et al., 2014; Pickhardt et al., 2003; Turaga et al., 2012). Goblet cell adenocarcinoma is seen in 19% of appendectomy specimens, while malignant NET represents 11% of malignant tumors^(Turaga et al., 2012).

Extremely difficult to diagnose prior to surgical inspection, and it usually depends on the pathology following appendectomy^(Deng et al., 2014). Appendix cancers may also be asymptomatic and be found incidentally as an abnormal appearing appendiceal orifice on colonoscopy, at surgery, or on cross-sectional imaging for other indications.

There are no established risk factors for the development of appendix cancer. Malignant appendix tumors most often present with acute appendicitis and diagnosed incidentally at histologic assessment of the surgical specimen.

Mucinous epithelial neoplasms secrete mucinous fluid, resulting in collections that are recognizable at cross-sectional imaging as mucocoele, periappendiceal or localized mucinous deposits, or PMP. Extraperitoneal dissemination of mucinous neoplasms is uncommon. PMP is a clinical syndrome characterized by the presence of mucin and/or epithelial cells within the peritoneum and on the serosa of the abdominal and pelvic organs^[12]. Nonmucinous epithelial tumors are similar histologically and radiologically to colon carcinomas,

manifesting as a focal mass with possible regional lymph node involvement and metastatic disease to the peritoneum, liver, or lung^(Pickhardt et al., 2003).

LAMNs have low-grade cytologic atypia. HAMNs have all the features of LAMN, but with high-grade cytologic atypia. Mucinous adenocarcinomas show infiltrative invasion and may be well, moderately, or poorly differentiated^(Carr et al., 2016).

Ultrasonography (US) is a valuable first pass tool for evaluation of the appendix, with sensitivity and specificity of 88% and 93%, respectively, for detection of appendicitis^(Parker et al., 2014). CT with or without intravenous and oral contrast material is used at many institutions as the initial imaging modality for evaluation of right lower quadrant or diffuse abdominal pain, which may lead to detection of appendiceal tumors^(Pickhardt et al., 2003).

Optimal treatment for epithelial mucinous neoplasms depends on the histologic characteristics of the primary tumor and peritoneal disease, as well as the TNM stage^(Kelly, 2015). The treatment for adenoma and LAMN confined to the appendix is appendectomy. Cases of LAMN with nodal involvement, HAMN, and adenocarcinoma without peritoneal extension are treated with appendectomy, right hemicolectomy, and lymph node dissection. Patients with peritoneal spread undergo laparoscopy, and based on the extent and histologic grade of the peritoneal tumor, CRS and HIPEC. Patients not eligible for complete CRS should undergo systemic chemotherapy with regimens used for metastatic colorectal cancer^(Kelly, 2015).

In patients with mucinous neoplasms and peritoneal spread of disease, a combined approach consisting of laparoscopy, CRS, and HIPEC is considered the standard of care^(Carr et al., 2016; Kelly, 2015; Sugarbaker, 2006). Laparoscopy plays an important role in management of patients with peritoneal spread. The addition of laparoscopy to preoperative CT, physical examination, and endoscopy improves the completion rates for CRS and HIPEC from 56% to 70%^(Iversen et al., 2013). Malignant tumors with T4a disease and high-grade histologic features are treated with laparoscopy and CRS—with removal of any residual disease after appendectomy, right hemicolectomy, omentectomy, and right lower quadrant peritonectomy—as well as HIPEC^(Kelly, 2015).

CONCLUSION:

Appendiceal tumors are rare with majority of appendiceal neoplasms comprised of epithelial neoplasms and NETs. Mostly present with abdominal pain, manifest as acute appendicitis, or are incidentally discovered in appendectomy specimens during histopathological evaluation. Imaging findings tend to be suggestive of acute appendicitis most of the times. CT is often used for staging of appendiceal tumors and subsequent follow up. In majority of cases, appendectomy will be sufficient, and some cases may require right hemicolectomy. Cytoreductive surgery (CRS) and intraperitoneal chemotherapy have improved the prognosis of patients with peritoneal disease.

REFERENCES

- Carr, N. J., Cecil, T. D., Mohamed, F., Sobin, L. H., Sugarbaker, P. H., González-Moreno, S., Taflampas, P., Chapman, S., Moran, B. J., & Peritoneal Surface Oncology Group International. (2016). A Consensus for Classification and Pathologic Reporting of Pseudomyxoma Peritonei and Associated Appendiceal Neoplasia: The Results of the Peritoneal Surface Oncology Group International (PSOGI) Modified Delphi Process. *The American Journal of Surgical Pathology*, 40(1), 14–26. <https://doi.org/10.1097/PAS.0000000000000535>
- Connor, S. J., Hanna, G. B., & Frizelle, F. A. (1998). Appendiceal tumors: Retrospective clinicopathologic analysis of appendiceal tumors from 7,970 appendectomies. *Diseases of the Colon and Rectum*, 41(1), 75–80. <https://doi.org/10.1007/BF02236899>
- Deng, K., Zhang, C.-Q., Wang, G.-L., & Li, W. (2014). Primary appendiceal mucinous adenocarcinoma mimicking bladder carcinoma: A case report and review of the literature. *Oncology Letters*, 7(4), 1270–1272. <https://doi.org/10.3892/ol.2014.1842>
- Deshmukh, S., Verde, F., Johnson, P. T., Fishman, E. K., & Macura, K. J. (2014). Anatomical variants and pathologies of the vermiform appendix. *Emergency Radiology*, 21(5), 543–552. <https://doi.org/10.1007/s10140-014-1206-4>
- Iversen, L. H., Rasmussen, P. C., & Laurberg, S. (2013). Value of laparoscopy before cytoreductive surgery and hyperthermic intraperitoneal chemotherapy for peritoneal carcinomatosis. *The British Journal of Surgery*, 100(2), 285–292. <https://doi.org/10.1002/bjs.8908>
- Kelly, K. J. (2015). Management of Appendix Cancer. *Clinics in Colon and Rectal Surgery*, 28(4), 247–255. <https://doi.org/10.1055/s-0035-1564433>
- Pickhardt, P. J., Levy, A. D., Rohrmann, C. A., & Kende, A. I. (2003). Primary neoplasms of the appendix: Radiologic spectrum of disease with pathologic correlation. *Radiographics: A Review Publication of the Radiological Society of North America, Inc.*, 23(3), 645–662. <https://doi.org/10.1148/rf.233025134>
- Sugarbaker, P. H. (2006). New standard of care for appendiceal epithelial neoplasms and pseudomyxoma peritonei syndrome? *The Lancet. Oncology*, 7(1), 69–76. [https://doi.org/10.1016/S1470-2045\(05\)70539-8](https://doi.org/10.1016/S1470-2045(05)70539-8)
- Turaga, K. K., Pappas, S. G., & Gamblin, T. C. (2012). Importance of histologic subtype in the staging of appendiceal tumors. *Annals of Surgical Oncology*, 19(5), 1379–1385. <https://doi.org/10.1245/s10434-012-2238-1>