Original Research Paper

Volume - 4 | Issue - 4 | July - 2020 | ISSN (O) : 2663-0729 | ISSN (P) : 2663-0710

CARCINOID APPENDIX PRESENTING AS ACUTE APPENDICITIS: A CASE REPORT

Ritesh Kumar Senior Resident, Department of General Surgery Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India.

Amit Jangra Junior Resident, Department of General Surgery Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India.

Naresh Pal Associate Professor, Department of General Surgery Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India.

ABSTRACT

Carcinoid or neuroendocrine tumor are the most common primary neoplasm encompassing for nearly 50% of the appendiceal tumors. Usually they are asymptomatic, however some patients may even present with acute appendicitis. These tumorsare discovered either incidentally during laparotomy or appendectomy done for treatment of patients with acute appendicitis. Surgical resection remains the mainstay in management of carcinoid tumor of appendix. Tumors which are less than 1 cm in size can be managed by appendectomy alone as they rarely metastasize. However for tumors greater than 2cm in size, there is an increase in the risk of metastasis and such cases are managed by right hemicolectomy. For tumors measuring 1-2 cm in size, the type of surgery will depend upon various factors. In this report we present a case of a 25 year male which underwent surgery for management of acute appendicitis which on histopathological examination was reported as carcinoid tumor.

KEYWORD

carcinoid, appendix, neuroendocrine tumour, appendiceal tumors, appendicectomy, right hemicolectomy

*Corresponding Author Ritesh Kumar

Senior Resident, Department of General Surgery Pt. B. D. Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India. dr.riteshkumar25@gmail.com

Introduction

Acute appendicitis is one of the most frequently encountered surgical emergencies. Out of which appendiceal neoplasms comprise nearly 0.5% of the appendectomies. Carcinoid appendix or neuroendocrine tumor are the most common primary tumor of appendix which encompasses for nearly 50% of the appendiceal neoplasms. Unlike the other appendiceal neoplasm, carcinoids are encountered in the younger age group with the highest incidence between the age group of 15-19 yrs in females and 20-29 years in males. These tumors are more frequently encountered in females than males. Carcinoid tumors of appendix are usually asymptomatic though in some patients they may present with appendicitis. Most of them are removed either incidentally during laparotomy or appendectomy done for management of patients with appendicitis. The most common site for carcinoidis the tip of appendix (60%-80%) followed by body (5%-21%) and base of the appendix (7%-10%). As compared to other appendiceal neoplasms, neuroendocrine tumors of appendix carry a favourable prognosis with 5 year survival rate upto 92%. Surgical resection remains the mainstay in management of these neoplasms with appendectomy alone suffices in most of the patients. We present a case of 25 year male who underwent emergency appendectomy for acute appendicitis which on histopathological examination was reported as carcinoid tumor.

Case Report

A 25 year male presented to the emergency with complaint of pain in right lower abdomen for 2 days. The pain was initially localised in the periumbilical region but later on it shifted to the right iliac fossa. Patient had anorexia however; there was no history of fever, vomiting, excessive sweating, diarrhoea or difficulty in breathing. The patient had a history of similar complaints in the past for which he was managed conservatively at a regional hospital. On general physical examination, thepatient was afebrile and vital parameters were within normal limits. Laboratory findings revealed the total leucocyte counts was 14000/mm3. Rest of the findings were within normal limits. On abdominal examination, there was tenderness and rebound tenderness in the right iliac fossa. Ultrasonography of abdomen revealed a blind ended tubular structure in the right iliac fossa with maximum diameter of 8mm suggestive of acute appendicitis. Patient was operated for emergency open appendectomy. Intraoperatively, tip of appendix was inflamed and base was healthy. Post-operatively, patient progressed well and was discharged on third day. Histopathology report showed marked inflammation in the wall of appendix with inflammatory exudates with area of carcinoid tumor at the tip of appendix (Fig 1a, 1b). It was confirmed by immunohistochemistry which was positive for synaptophysin, chromogranin and neuron specific enolase (Fig 2a, 2b, 2c).

Figure 1a, 1b

H&E sections showing islands of tumor cells involving the tip of appendix (black arrows)

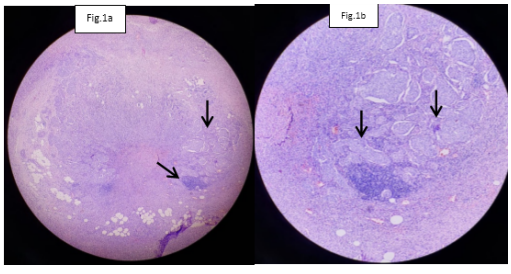
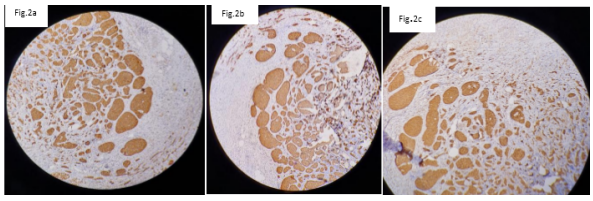


Figure 2

Immunohistochemistry showing tumor cells



Note. Immunohistochemistry showing tumor cells which are positive for synaptophysin, chromogranin and neuron specific enolase in figure 2a, 2b, 2c respectively.

Discussion

Carcinoid tumors are derived from the enterochromaffin cells of gastrointestinal tract. In the appendix they arise from the subepithelium and have an indolent behaviour. Appendix is the second most common site of carcinoid tumor after small bowel. Carcinoids are frequently encountered on tip of appendix followed by body and base. These tumors have a female preponderance with most patients presenting in the younger age group.

Appendiceal carcinoids are benign lesions which are usually asymptomatic and may present with acute appendicitis due to the obstruction of appendiceal lumen. Symptoms of classic carcinoid syndrome such as flushing, diarrhoea and cardiac disease are observed only if the tumor is of large size or has distant metastasis. Appendiceal tumors are rare and are incidentally detected during laprotomy or appendectomy. Carcinoid tumors account for nearly 50% of appendiceal neoplasms. In the present case, carcinoid tumor involved the tip of appendix and presented with typical history and findings of acute appendicitis which was confirmed on histopathology. However luminal obstruction was not the cause as the tumor was present at the tip of the appendix. The histopathological diagnosis was confirmed by immunohistochemistry which showed positivity for neuroendocrine markers such as synaptophysin, chromogranin and neuron specific enolase.

Surgical resection is the main treatment modality for the carcinoid tumor of the appendix. Their malignant potential and the extent of resection depend on the size of the tumor. For the lesions smaller than 1 cm appendectomy alone is the sufficient as they rarely metastasize. Whereas tumors of more than 2 cm in size have an increased risk of metastasis and require right hemicolectomy. However for tumors ranging between 1-2 cm, the type of surgery will depend on various factors such as high grade, positive surgical margin, mesoappendiceal extension of tumor and presence of tumor at the base of appendix. Tumors with these factors require right hemicolectomy otherwise appendicectomy alone can be done. In the reported case the tumor was less than 1 cm in size and was located at the tip of appendix with clear surgical margins hence appendicectomy was preferred modality. Long term follow up of the patients is required if there is lymph node involvement or when the tumor is high grade. Tumors of size 1-2 cm require follow up only if high risk factors are present. For tumors measuring less than 1 cm and 1-2 cm in size do not require any follow up if risk factors are not present. Patients with appendiceal carcinoid have high incidence of synchronous and metachronous gastrointestinal lesions thus requiring close follow up and screening. Prognosis of appendiceal carcinoid tumors is excellent with a 5 year survival rate of 92% in the patients with local disease, 81% in patients with regional metastasis and 31%in patients with distant metastasis.

Conclusion

Appendicitis is the most commonly encountered emergency by a general surgeon. Carcinoid tumor is usually asymptomatic but some patients may present with acute appendicitis. Hence possibility must always be kept in mind. Once the diagnosis of carcinoid tumor is made, appropriate staging and treatment should be done. Tumors greater than 2 cm and 1-2 cm in size with high risk factors usually require right hemicolectomy while appendicectomy is sufficient for tumors of small size with no risk factors. Patients with appendiceal carcinoid have high incidence of synchronous and metachronous gastrointestinal tumour.

References

1. Co kun H, Bostanci O, Dilege ME, et al. (2006). Carcinoid tumors of appendix: treatment and outcome. Ulus Travma Acil Cerrahi Derg, 12, 150–154.
2. Deans GT and Spence RA. (1995). Neoplastic lesions of the appendix. Br J Surg, 82, 299-306.
3. Gu Y, Wang N, Xu H. (2015). Carcinoid tumor of the appendix: A case report. Oncology letters, 9, 2401-2403.
4. Hemminki K, Li X. (2007). Incidence trends and risk factors of carcinoid tumors: a nationwide epidemiologic study from Sweden. Cancer, 92(8), 2204-10.
5. MacGillivray DC, Heaton RB, Rushin JM, Cruess DF. (1992). Distant metastasis from a carcinoid tumor of the appendix less than one centimeter in size. Surgery, 111, 466–471.
6. Modlin IM, Lye KD and Kidd M. (2003). A 5-decade analysis of 13,715 carcinoid tumors. Cancer, 97, 934-959.
7. Moertel CG, Weiland LH, Nagorney DM, Dockerty MB. (1987). Carcinoid tumor of the appendix: treatment and prognosis. The New England Journal of Medicine, 317(27), 1699-1701.
8. Morais C, Silva E, Brandao PN et al. (2019). Neuroendocrine tumor of the appendix- a case report and review of the literature. Journal of Surgical Case Reports, 3, 1-4.
9. Neto IJF, Lopes AE, MarquesRD et al. (2014). Carcinoid tumor of cecal appendix: one-year incidence at Santa Marcelina Hospital. J Coloproctol (Rio J), 34(4), 245-249.
10. Pawa N, Clift AK, Osmani H, Drymousis P, Cichocki A, Flora R, et al. (2018). Surgical management of patients with neuroendocrine neoplasms of the appendix: appendectomy or more. Neuroendocrinology, 106, 242–251.
11. Pickhardt PJ, Levy AD, Rohrmann CA Jr and Kende AI. (2002). Primary neoplasms of the appendix manifesting as acute appendicitis: CT findings with pathologic comparison. Radiology, 224, 775-781.
12. Quaedvlieg PF, Visser O, Lamers CB, et al. (2001). Epidemiology and survival in patients with carcinoid disease in The Netherlands. An epidemiological study with 2391 patients. Ann Oncol, 12, 1295–1300.
13. Roggo A, Wood WC & Ottinger LW. (1993). Carcinoid tumors of the appendix. Annals of Surgery, 217(4), 385–390.
14. Ruffolo C, Fiorot A, Pagura G et al. (2013). Acute appendicitis: what is the gold standard of treatment? World J Gastroenterol, 19, 8799-8807.