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Case Report

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Carcinoid tumour of appendix

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ABSTRACT

Carcinoid tumors are quite common neoplasm of the appendix. The clinical presentation of these lesions somewhat corelates to that of acute appendicitis, or the tumors are asymptomatic. The carcinoids are commonly found incidentally during histopathological examination of the resected appendix following appendectomy or other abdominal procedures. Appendiceal carcinoids usually behave as benign tumors and appendicectomy alone is a sufficient treatment in the majority of cases, while for larger lesions (lesions at the base of the appendix), right colectomy should be performed. The prognosis of patients with local appendiceal carcinoids is excellent. The present study reports the case of a 49-years-old male patient that presented with recurrent abdominal pain of 6 weeks duration. The patient underwent successful appendectomy and recovered four days later. Subsequently, an appendiceal carcinoid tumor located at the tip of the appendix was diagnosed by histopathological examination. Follow-up examination one year after surgery revealed that the patient was well with no discomfort.

Keywords: Carcinoid tumor, Appendix, Appendicectomy

INTRODUCTION

Inspite of their low incidence's carcinoid tumors are constantly present in clinical practice. These neoplasms are neuroectodermal in origin and are classified under APUD neoplasms (amine precursor uptake and decarboxylation). In 1907 Oberndorfer first coined the term carcinoid, meaning 'cancer-like', to describe a rare ileal tumor with less malignant behavior than the more commonly identified large bowel carcinomas. Now, it is clear that carcinoids are, indeed, malignant. They are derived from the diffuse neuroendocrine system that is composed of peptide- and amine-producing cells that may secrete different hormones depending on the site of origin.[1] The WHO recently dropped the term carcinoid and now classifies these tumors as NETs, although carcinoid may likely remain in the vernacular.

Carcinoid tumors constitute an important group of neoplasms requiring careful assessment and discerning management. While, carcinoid tumors of the small intestine are most common cause of clinical symptoms, carcnoid tumor of appendix are almost always found incidentally during surgery for appendicitis and other surgical procedures. Appendiceal carcinoid tumors behave in a benign manner and metastasize in less than 2%. This occurs in tumor more than 2 cm in diameter.

The case report reviewed our own experience of appendix carcinoid tumor treated at Kalinga Institute of Medical Sciences, Bhubaneswar.

CASE REPORT

A 49-years-old patient was admitted at Kalinga Institute of Medical Sciences, Bhubaneswar as a follow up case of appendicular abscess with chief complaints of pain in whole abdomen since last 6 weeks and 2 episodes of vomiting in last 3 days. The patient reported a 6 weeks history of recurrent dull whole abdominal pain that had not

been considered serious enough to obtain medical assistance. Three days prior to admittance, the aforementioned symptoms became aggravated and the patient presented to the Kalinga institute of medical sciences. Physical examination revealed a body temperature of 37°C, blood pressure of 136/78 mmHg and a pulse rate of 105 beats/min. During assessment, the patient experienced tenderness on superficial palpation, but no rebound tenderness, in the right lower abdomen. No palpable masses were observed in the abdomen. Laboratory tests were performed, yielding the following results: white blood cell count, 7×109 cells/l (normal range, 4-10×109 cells/l); neutrophil proportion, 70% (normal range, 45-80%); hemoglobin level, 13 g/dl (normal range, 11-15 g/dl); platelet count, 190×109 platelets/l (normal range, 100-300×109 platelets/l).

In addition, abdominal computed tomography revealed mild focal dilated appendix with surrounding fat stranding. The patient possessed no previous medical history and no family medical history of appendicitis.

The patient underwent surgery for the treatment of the acute attack of chronic appendicitis. The appendix was identified as 4 cm long and 0.6 cm in diameter, demonstrating hyperemia and edema and lumen containing fecolith. Outer surface appeared smooth dull and congested focally. The resection of the appendix was completed without complications and the patient recovered four days later. The specimen was examined by physiological analysis and stained using hematoxylin and eosin. At the tip of the appendix, there was a well-differentiated tumor, measuring 2.5 mm at the maximum dimension confined to the submucosa of the tip of the appendix.

The mitotic activity of the lesion was not notable. The diagnosis of appendiceal carcinoid tumor was confirmed by positive immunostaining for the neuroendocrine marker's chromogranin A and synaptophysin. No further treatment was administered. Follow-up examination after surgery revealed that the patient was well, with no discomfort.

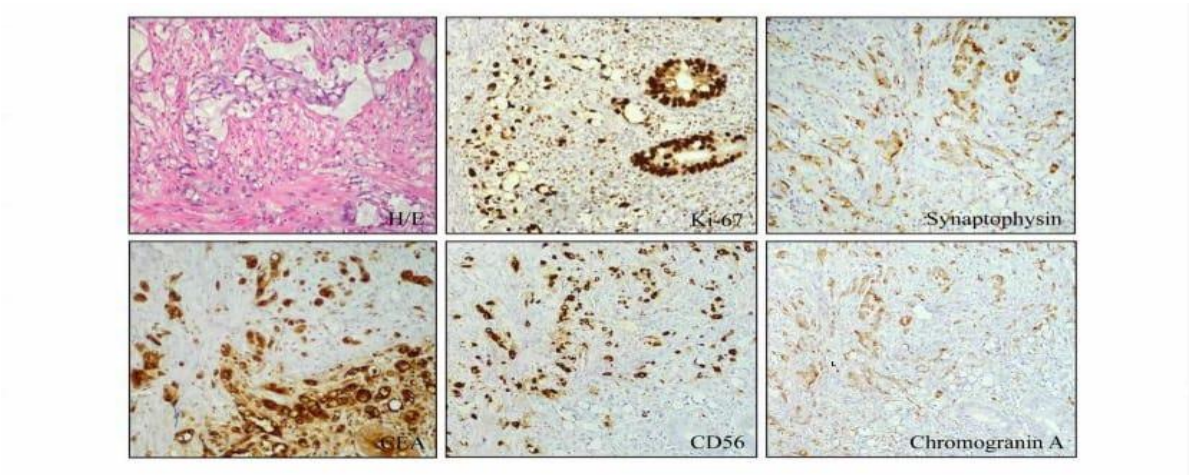

Figure 1: Slides stained with tissue markers.

DISCUSSION

Carcinoid tumors of the appendix are relatively uncommon neoplasms. It has been previously reported that the incidence of appendiceal carcinoids ranges between 0.3 and 0.9%, as determined using histopathological examination performed on excised appendectomy specimens.[2,3] However, appendiceal carcinoids are the most frequent tumors arising from the appendix, comprising between 32 and 57% of all appendiceal tumors.[4] Carcinoid tumors are neoplasms derived from the subepithelial neuroendocrine cells of the appendix, that rarely cause metastatic disease.[5] There is no specific pre-operative clinical presentation for appendiceal carcinoids. In general, appendiceal carcinoids are either asymptomatic or present as acute appendicitis, which is then diagnosed incidentally as appendiceal carcinoids during surgery.[6] In addition, the carcinoids can result in recurrent episodes of abdominal pain due to partial obstruction of the appendiceal lumen by a tumor.

The presence of neuroendocrine symptoms, including flushing, diarrhea and cardiac disease, are rarely reported.[7] In the case of the present patient, the tumor was located at the tip of the appendix, and the patient experienced recurrent right lower abdominal pain that had been present for three years. The surgical procedure was performed for the treatment of appendicitis, and the carcinoid tumor was identified incidentally during the histological examination of the excised surgical specimen.

Appendiceal carcinoids grow slowly, and the overall prognosis is excellent.[8] At present, tumor size is the most reliable indicator for the assessment of the malignant potential of lesions. In the majority of patients with appendiceal carcinoids, the tumor diameter is 2 cm in diameter. This follow-up procedure comprises a history and physical examination every 3-12 months post-resection and every 6-12 months thereafter for ≤10 years, with consideration of follow-up imaging or laboratory

markers, such as 5-hydroxyindoleacetic acid or chromogranin A.[11]

Overall, the long-term prognosis of appendiceal carcinoids is extremely good. Sandor and Modlin evaluated 1570 carcinoid tumors of the appendix that were treated at the American National Cancer Institute between 1973 and 1991. It was reported that the five-year survival rates of patients with localized, regional metastases and distant metastases in appendiceal carcinoids were 94.0, 84.6 and 33.7%, respectively, and the overall five-year survival rate was 85.9%.[4] Another study with a large cohort performed by Modlin et al reported that patients with local disease demonstrate a five-years survival rate of 92%, those with regional metastases demonstrated a five-years survival rate of 81% and the few with distant metastases demonstrated a five-year survival rate of 31%.[5] Carcinoid tumors are the most common neoplasms of the appendix, but demonstrate no specific clinical presentation. The presence of these tumors should be considered, particularly during surgical procedure.

CONCLUSION

Appendix carcinoid tumors are quite rare, usually asymptomatic and diagnosed incidentally on histopathological examination after appendectomy. The treatment of carcinoid tumors of the appendix is directly related to the tumor size, localization, presence of lymphovascular and mesoappendix invasion, mitotic activation rate and level of Ki67. Thus, it is important to follow the histopathological results after appendectomy. The prognosis of appendix carcinoid tumors is very good if the appendix is non-perforated.

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