

Incidental Appendiceal Carcinoids: A Rare Case at an Uncommon Site

Sujan Sharma^{1*}, Ramesh Makaju¹, Nripesh Rajbhandari²

We report a case of 10-year-old female, who was admitted in the surgical department of Dhulikhel Hospital with symptoms of acute appendicitis. The appendectomy was performed. The gross appearance of the appendix was of a slight catarrhal appendicitis, with no sign of a tumor, but the tip of the appendix was perforated. The histopathological examination revealed a carcinoid tumor on the tip of the appendix. No vascular or perineural invasion was seen. Conclusions: The appendectomy is enough for carcinoid tumors of the tip of the appendix, of small sizes, without vascular or perineural invasion.

KEYWORDS: young patient, acute appendicitis, carcinoid appendiceal tumor.

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Introduction

Carcinoid tumours of the appendix are uncommonly incidentally detected neuroendocrine neoplasms during histopathological examination following appendicectomy for acute appendicitis and is considered the most common type of appendiceal primary malignant lesion which are found in 0.3%-0.9% of patients undergoing appendicectomy [1] This tumor rarely presents with metastases. They are less common in children, with a reported incidence of 0.08 % [2]. We report here a case of carcinoid tumour of the tip of appendix in a young female child which was diagnosed by histopathological examination.

Case report

A 10-year-old female who presented with abdominal pain of two-day duration. She gave history of mild, intermittent abdominal pain over perumbilical region then shifted to right iliac fossa, fever was also acute on set followed by nausea. On examination per abdomen it was soft and tender. White blood count was 11,500 with 86% granulocytes, hemoglobin 12.5 gm% and platelets 3,18,000/ μ L. Ultrasonography findings likely of appendicitis with appendicular lump and abscess formation. The patient with all these findings was admitted in surgery ward. The emergency laparoscopic appendectomy was done.

Pathology

The serosal surface of the appendix was congested and appendix was sent in multiple bits, tip of the appendix was perforated. On cut section of the other bits lumen identified and filled with fecal matter. Section from the tip and other representative sections were given. Microscopic examination revealed a carcinoid tumor composed of solid nests and acini demonstrating uniform cells with a speckled nuclear chromatin pattern. Mitoses were rare and tumor involved all layers of the appendiceal wall and extended to the overlying serosal surface. Neutrophil was present within periappendiceal tissue.

Discussion

Carcinoid tumors were first described by a Swiss pathologist Theodor Langhans in 1867. These neoplasms are rare and of neuro-endocrine origin but is the most common tumor of the appendix, occurring in 0.226% of appendectomies performed at all ages [3]. Carcinoid of the appendix encountered in 1:100,000 to 169:100,000 children with female preponderance and median age of presentation is 12.3–13 year [4-6].

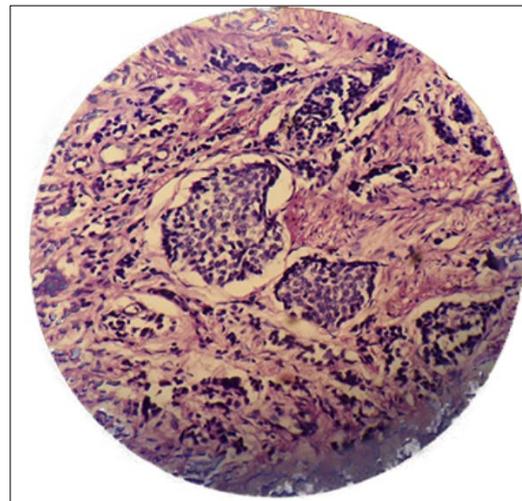


Figure 1. Section showing nest of tumor cells with speckled nuclear chromatin.

The location of the appendix was described by Beger in 1882 and the term was introduced by a

¹Department of Pathology, ²Department of Surgery, Kathmandu University School of Medical Sciences, Dhulikhel, Nepal.

pathologist, Siegfried Oberndorfer in 1907 [3]. Parkes *et al.*'s study over a period of 30 years, showed an incidence rate of 1.14 per 1 million children per year [7]. Clinically, it presents as acute appendicitis and sometimes as chronic abdominal pain. The most of the cases location of tumor is at the apex of the appendix followed by mid portion and the base [6]. Study done by Pelizzo *et al* reports incidence of carcinoid tumors located at the tip of the appendix and with size less than 10 mm present as acute appendicitis, while tumors which measures more than 20 mm with location at the base of the appendix may present as a case of peritonitis [8]. The predilection for benign carcinoids in younger patients (20-30 years of age) and their preferential location in the tip of the appendix are well recognised and is attributed to the origin of the tumor from subepithelial neuroendocrine cells [9]. Subepithelial neuroendocrine cells are more numerous at the tip of the appendix than at the base with increase of the age their density increases and the peak around the third decade of life followed by another decline with increase of age. Clinical symptoms of carcinoid

syndrome flushing, diarrhea, and wheezing are usually not found except in large tumor mass or distant metastasis [9, 10].

Literatures on the incidence of carcinoid tumors of appendix on young patients remains contradictory with some studies showing increase of incidence while other reports decrease in the prevalence of these tumors. These findings could be postulated to be linked to the number of appendectomies performed.

Conclusion

Carcinoid tumors are rare, but are the most common neuroendocrine neoplasms of the appendix which are incidentally detected during histopathological examination. The present case highlights the continuing need for histopathological examination of appendix after every appendectomy.

Conflict of Interest: None declared

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