

A Carcinoid Tumor of the Appendix in a Child: A Case Report

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Abstract

The appendix is one of the most common sites for carcinoid tumors. Most carcinoids are found in appendices removed incidentally at laparotomy for conditions unrelated to acute appendicitis. We describe the case of a 13-year-old female who presented with abdominal pain in the right lower quadrant (RLQ), with nausea and decreased appetite for the previous 2 days. A physical examination favored a diagnosis of acute appendicitis. A carcinoid tumor was diagnosed based on the histological examination of the removed appendix. The patient underwent an isolated appendectomy due to the small size of the lesion.

Key Words: Carcinoid tumor, Case report, Children.

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1- INTRODUCTION

The appendix is one of the most common sites for carcinoid tumors (1). Carcinoid tumors occur most commonly between the third and fourth decades. Carcinoid tumors are less common among children, with an incidence of 0.2–0.5% among resected surgical specimens (2). Most carcinoids are found in appendices removed incidentally at laparotomy for conditions unrelated to acute appendicitis or in specimens removed in cases of acute appendicitis. Usually, the tumors remain small and rarely metastasize. Therefore, they do not give rise to carcinoid syndrome (3).

The majority of carcinoids occur close to the tip of the appendix and have a yellow color after formalin fixation. They are typically round and well circumscribed, but diffuse forms can occur (4). Microscopically classic (insular) carcinoid tumors feature solid nests of small monotonous cells, with occasional acinar or rosette formation. Mitosis is extremely rare (5). We present a 13-year-old female child with carcinoid tumor of the appendix.

2- CASE REPORT

A 13-year-old female presented with abdominal pain in the right lower quadrant (RLQ), with nausea and decreased appetite for the previous 2 days. A physical examination favored a diagnosis of acute appendicitis. The patient underwent an emergency appendectomy. On the tip of the appendix, a firm yellowish mass with a diameter of 0.8 cm was identified (**Figure. 1**). The histological examination revealed a carcinoid tumor composed of solid nests and islets. Mitotic activity was insignificant. The tumor involved the muscular layer and extended to the overlying serosal surface. The tumor did not extend to the base of the appendix. **Figure.2** shows a photomicrograph of a typical appendiceal carcinoid tumor (Hematoxylin and Eosin, 40×), characterized by solid nests (islands) containing a uniform population of round to oval cells. In the present case, the cells had scant eosinophilic cytoplasm, with fine nuclear chromatin. Mitosis was rare (**Figure. 3**). The tumor cells invaded the muscularis layer and serosa.



Fig.1: Macroscopic picture of carcinoid tumor in present patient (hematoxylin and eosin, ×400).

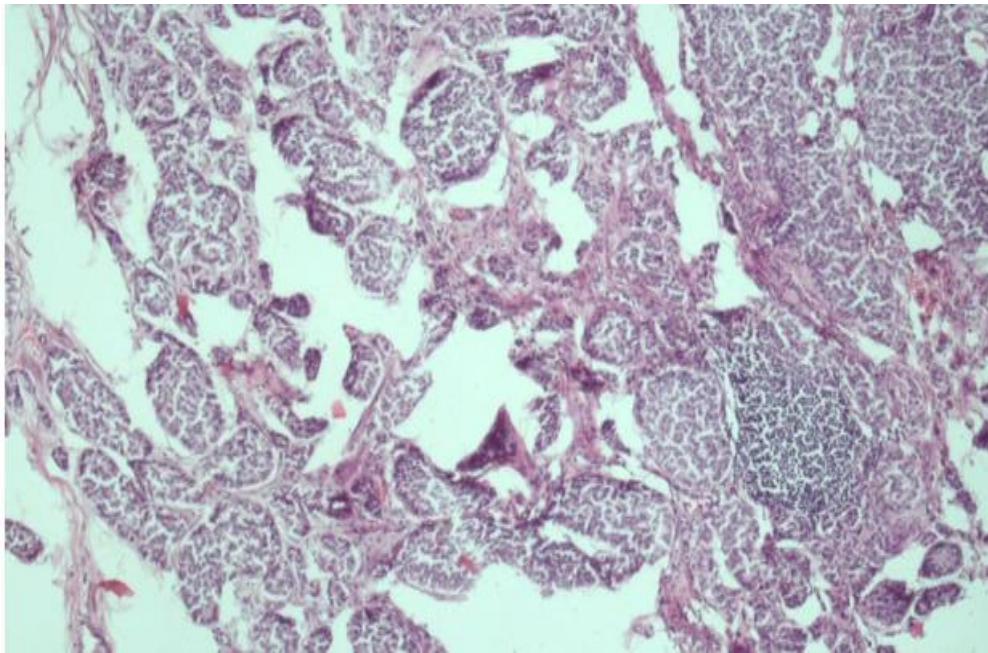


Fig.2: Solid nest of small monotonous cells (hematoxylin and eosin, $\times 400$).

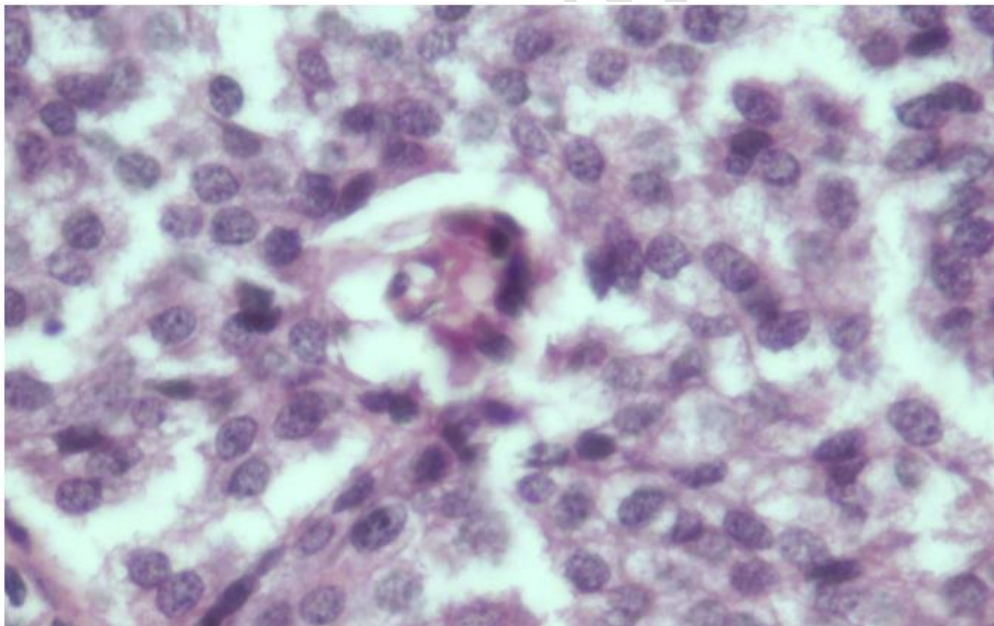


Fig.3: Monotonous cells with few eosinophilic cytoplasm and salt and pepper nuclei (hematoxylin and eosin, $\times 400$).

3- DISCUSSION

Carcinoid tumors of the appendix are relatively uncommon neoplasms. Although they are considered a rare pathology in children, they are the most frequent tumors of the gastrointestinal tract in childhood and adolescence (6). The clinical presentation of a carcinoid tumor of the appendix is similar to that of acute appendicitis, but it can be an incidental intraoperative discovery during appendectomy or other surgical procedures. Clinical symptoms of carcinoid syndrome include flushing, diarrhea, and wheezing, but these are usually not present, except in the case of a large tumor mass or distant metastasis (7).

The size and depth of invasion are important prognostic criteria, and tumors larger than 2 cm metastasize more frequently than smaller ones. The treatment of carcinoid tumors of the appendix depends on the size and site of the tumor. Tumors smaller than 2 cm would be resected when the mesoappendix is involved or in cases with residual tumor at the margin (8). The patient in the present case had an appendiceal carcinoid tumor, with a diameter of 0.8 cm and tumor-free margin. Therefore, simple appendectomy was considered adequate treatment for this patient.

4- CONCLUSION

Carcinoid tumors of the appendix in children are rare and under reported tumors. They are discovered incidentally, as most patients usually present with symptoms of acute appendicitis. Localised disease has an excellent prognosis, hence the clinicians should be aware of the tumor, which enables an early removal of the appendix when the probability of metastasis is still low, thus preventing the morbidity and mortality associated with advanced disease. The present patient underwent an isolated appendectomy due to the small size of the lesion.

5- CONFLICT OF INTEREST

The authors declare that there is no conflict of interests regarding the publication of this paper.

6- ACKNOWLEDGMENT

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7- REFERENCES

1. Tchana-Sato V, Detry O, Polus M, Thiry A, Detroz B, Maweja S, et al. Carcinoid tumor of the appendix: a consecutive series from 1237 appendectomies. *World Journal of Gastroenterol* 2006; 12(41): 6699-701.
2. Christianakis E, Paschalidis N, Chorti M, Filippou G, Rizos S, Filippou D. Carcinoid tumor of the appendix in children: a case report. *Cancer Journal* 2008; 1(1): 136.
3. Moertel CG, Weiland LH, Nagorney DM, Dockerty MB. Carcinoid tumor of the appendix: treatment and prognosis. *The New England Journal of Medicine* 1987; 317(27): 1699-701.
4. Lundqvist M, Wilander E. A study of the histopathogenesis of carcinoid tumors of the small intestine and appendix. *Cancer* 1987; 60(2): 201-6.
5. Soga J, Tazawa K. Pathologic analysis of carcinoids. Histologic evaluation of 62 cases. *Cancer* 1971; 28(4): 990-8.
6. Moertel CL, Weiland LH, Telander RL. Carcinoid tumor of the appendix in the first two decades of life. *Journal of Pediatric Surgery* 1990; 25(10): 1073-5.
7. Hemminki K, Li X. Incidence trends and risk factors of carcinoid tumors: a nationwide epidemiologic study from Sweden. *Cancer* 2007; 92(8): 2204-10.
8. D'Aleo C, Lazzareschi I, Ruggiero A, Riccardi R. Riccardi Carcinoid tumors of the appendix in children: two case reports and review of the literature. *Pediatric Hematology and Oncology* 2001; 18(5): 347-51.