Research Paper

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Carcinoid Tumour of Appendix in 18 Year Old Boy: A Rare Case Report

MEDICAL SCIENCE

KEYWORDS : Carcinoid, Appendix.

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ABSTRACT

Carcinoid tumours are the rare neuroendocrine neoplasms, occurring most commonly in the appendix. The clinical presentation of the appendiceal carcinoids is similar to that of acute appendicitis, although in many cases the tumour is diagnosed incidentally. The diagnosis should be confirmed histologically. Localized disease has an excellent prognosis, and the tumor usually has little metastatic potential. We report a case of a carcinoid tumour in the body of the appendix of a eighteen year old boy which was diagnosed incidentally on histopathological examination.

INTRODUCTION

Carcinoid tumours are rare neuroendocrine neoplasms, occurring with higher frequency in the appendix. Histopathologically, appendiceal carcinoid tumor is mostly enterochromaffin (EC) cell type and derives from a subepithelial cell population, which is different from neuroendocrine tumor in other sites [1]. Although rare and usually detected incidentally in appendicectomy, it is considered the most common type of appendiceal primary malignant lesion, and is found in 0.3%-0.9% of patients undergoing appendicectomy[2]. This tumor rarely presents with metastases [3]. They are less common in children, with a reported incidence of 0.08% [4]. We report here a case of carcinoid tumour of the body of appendix in a young boy which was diagnosed by histopathological examination.

CASE REPORT

The patient is 18-year-old male who presented with severe right lower quadrant abdominal pain of three days duration. He gave history of mild, intermittent abdominal pain for a few weeks. There was no history of flushing, diarrhea, vomiting, urinary symptoms, food allergies, weight loss or fever. His family history was not significant.

Physical examination revealed a healthy looking male with mild distress, a temperature of 101.2 degrees and blood pressure 120/80. He had severe direct and rebound tenderness in the right lower quadrant. He had no hepatospenomegaly or lymphadenopathy and the rest of his physical examination was normal.

White blood cell count was 16,200/uL consisting of predominantly neutrophils, hemoglobin 13.8 gm%, hematocrit 41%, and platelets 189,000/uL. Serum glucose was 105 mg%, urea nitrogen 12 mg%, creatinine 0.8 mg% and electrolytes within normal range. Computerized tomography (CT) of the abdomen was consistent with appendicitis, the rest of the abdominal and pelvic organs were normal. He underwent appendicectomy and the specimen was sent for histopathological examination.

PATHOLOGY

Gross: The appendix was enlarged in size and the serosal surface was covered with a fibrinopurulent exudate. The lumen was dilated and 1.5 cm, circumscribed, nonencapsulated, firm, tan-yellow nodule was identified within the body of the appendix, located at a distance of 0.7 cm from the surgical resection margin.

Microscopic examination revealed a carcinoid tumor composed of solid nests and acini demonstrating uniform cells with a speckled nuclear chromatin pattern (Figure 1). Mitoses was minimal and mucinous cells were identified. Tumor involved all the layers of the appendiceal wall and extended to the overlying serosal surface. The base of the appendix was free of tumor.

Immunohistological analysis demonstrated positive staining of tumor cells for neuroendocrine markers including Chromogranin and Synaptophysin (Figure 2&3). An inflammatory cell infiltrate predominantly composed of neutrophils was found within the lumen of the appendix and extensively within periappendiceal tissue.

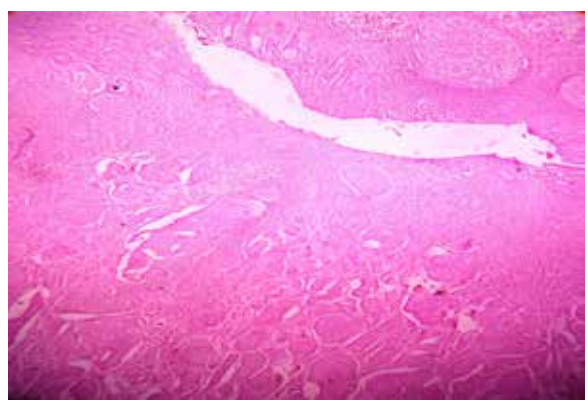

Figure 1: Photomicroscopy showing solid nests, cords and islands of tumour cells in mucosa and submucosa. (Hematoxylin and eosin stain –10×)

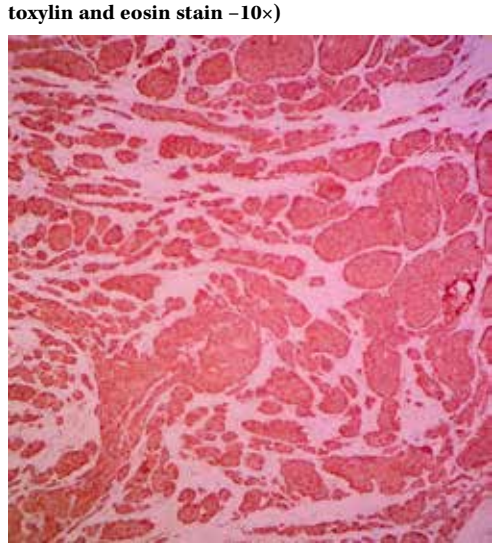

Figure 2: Chromogranin positivity in the

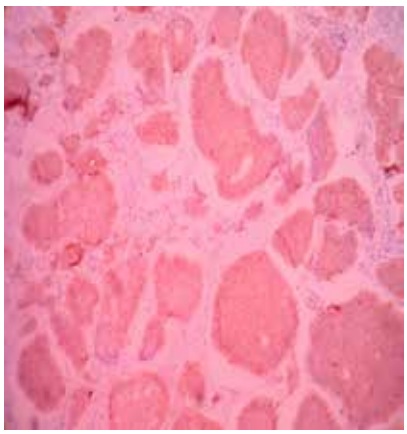

Figure 3: Synaptophysin stain demonstrates

cytoplasm of tumour cells (40×) positive staining of tumor cells

DISCUSSION

First recognized in 1867, carcinoid tumors are rare malignant neuro-endocrine tumors. It is the most common tumor of the appendix, occurring in 0.226% of appendectomies performed at all ages [5]. The tumor is more common in white female children with a median age of 12.3–13 years [6–8], but it has been reported as early as three years of age.

Acute appendicitis is more common in pre-teen and teen-age boys, while female to male occurrence in carcinoid of the appendix is 2:1 [7]. Most carcinoid tumors in children arise in the appendix; however, they can also occur in other primary sites including the small intestine, bronchus and others [6].

Clinical presentation of carcinoid of the appendix is similar to acute appendicitis, but it can be an incidental intra-operative finding during appendicectomy or other surgical procedures. The majority of tumors are discovered on histological examination of the surgical specimen. Clinical symptoms of carcinoid syndrome including flushing, diarrhea, and wheezing are usually not found except in large tumor mass or distant metastasis [5]. This malignancy should be considered in the differential diagnosis of children, presenting with right lower quadrant pain, persistent or recurrent pneumonia or symptoms of carcinoid syndrome.

The tumor is localized at the apex of the appendix in 75% of cases, in the mid portion in 20% and in the following base in 5% [7]. Serum chromogranin A is a useful immuno-histo- chemical marker which is reliable in the diagnosis of various endocrine tumors including carcinoid.

Out of 1500 appendicectomies done at our institute, only the present case of carcinoid tumour of appendix has been detected resulting in the frequency of 0.06%.

CONCLUSION

Carcinoid tumors are rare, but are the most common tumors of the appendix. The clinical presentation of the carcinoid tumor of appendix is similar to acute appendicitis, but it remains an incidental diagnosis. The present case highlights the continuing need for histopathological examination of appendix after every appendicectomy.

REFERENCE

1. Stinner B, Rothmund M. Neuroendocrine tumours (carcinoids) of the appendix. Best Pract Res Clin Gastroenterol 2005; 19: 729-738 | 2. Goede AC, Caplin ME, Winslet MC. Carcinoid tumour of the appendix. Br J Surg 2003; 90: 1317-132 | 3. Roggo A, Wood WC, Ottinger LW. Carcinoid tumors of the appendix. Ann Surg 1993; 217: 385-390 | 4. Dall'Igna P, Ferrari A, Luzzatto C, Bisogno G, Casanova M, Alaggio R, et al. Carcinoid tumour of the appendix in childhood: The experience of two Italian Institutions. J Pediatr Gastroenterol Nutr 2005;40:216-9. | 5. Doede T, Foss HD, Waldschmidt J: Carcinoid tumors of the appendix in children—epidemiology, clinical aspects and procedure. Eur J Pediatr Surg, 2000; 10: 372-7 | 6. Spunt SL, Pratt CB, Rao BN et al: Childhood carcinoid tumors: the St Jude Children's research hospital experience. J Pediatr Surg, 2000; 35: 1282-6 | 7. Prommegger R, Obrist P, Ensinger C et al: Retrospective evaluation of carcinoid tumors of the appendix in children. World J Surg, 2002; 26:1489-92 | 8. Pelizzo G, La Riccia A, Bouvier R, Chappuis JP, Franchella A: Carcinoid tumors of the appendix in children. Pediatr Surg Int, 2001;17: 399-402 |