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CASE REPORT

An Unusual Case of Appendiceal Carcinoid Tumor in a Child-Case Report

S. Sushma[1] · **C. S. B. R. Prasad**[1] · **K. Mohan Kumar**[2]

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Abstract Carcinoid tumors of the appendix are rare in children. They clinically present with features of acute appendicitis and are often diagnosed incidentally on the resected specimens. Local disease carries an excellent prognosis and simple appendicectomy suffices, whereas advanced cases need a right hemicolectomy. Clinicians must be aware of this under reported and rare tumor in chidren. We report a case of appendiceal carcinoid tumor in a 10 year old male child who presented with clinical features of acute appendicitis which was diagnosed on histopathological examination.

Keywords Carcinoid tumor · Appendicectomy · Child

Introduction

Carcinoid tumors of the appendix in children are rare and incidence is of about one case per thousand evaluated children [1]. Though the clinical presentation of this tumors is similar to acute appendicitis, often they are incidentally diagnosed on histological examination of the resected surgical specimen [2]. In chidren, tumours are usually smaller than 2 cms in diameter and behave in benign fashion, although some have a potential for malignancy and metastasis [3]. In most of the cases, a simple appendicectomy is sufficient, however a right hemicolectomy maybe necessary in others.

Herein, we report a case of carcinoid tumour of appendix in a 10 year child which was detected incidentally on histopathological examination of the specimen resected for acute appendicitis. This emphasizes on the fact that a differential diagnosis of carcinoid tumor should be entertained in a child presenting as acute appendicitis, enabling an early diagnosis and treatment.

Case Report

A 10 year old male child presented with pain in the right iliac fossa since 1 day. There was no history of nausea, vomiting, flushing, diarrhea, urinary symptoms, weight loss or fever. Physical examination revealed a healthy looking male child in mild distress, pulse was 82b/min, B.P- 110/70 mmHg. He had direct and rebound tenderness in the right lower abdominal quadrant. No organomegaly/ lymphadenopathy was noted.

Investigations- Complete blood counts, Liver function tests, renal function tests, serum electrolytes were normal. Abdominal ultrasound revealed an acutely inflamed appendix with no free fluid in the abdomen.

The patient underwent a laproscopic appendicectomy. The resected specimen was 10 cm in length. External surface showed congested blood vessels. Cut surface showed a circumscribed, solid, firm yellow mass in the tip of the appendix with a diameter of 13 mm. Tumor was 8cms away from the surgical resected margin. (Fig. 1)

Microscopy revealed a tumor composed of solid nests of small, monotonous tumor cells with speckled nuclear

✉ S. Sushma
dr.sushma1985@gmail.com

[1] Department of Pathology, Sri Devaraj Urs Medical College, SDUAHER, Tamaka, Kolar, Karnataka, India

[2] Department of Surgery, Sri Devaraj Urs Medical College, SDUAHER, Tamaka, Kolar, Karnataka, India

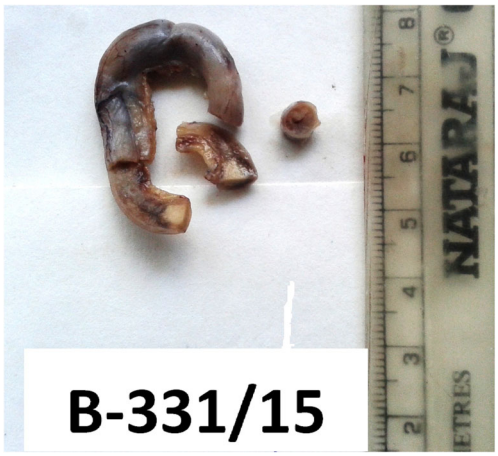

Fig. 1 Gross photograph showing a well circumscribed grey yellow tumor in the tip of appendix

chromatin pattern and scant cytoplasm, surrounded by clear retraction spaces. These tumour nests were seen infiltrating all the layers of appendiceal wall, extending into the mesoappendix. Base of the appendix was free from tumour. Mitotic rate of 1/10Hpf was noted. There was no evidence of lymphovascular/ perineural invasion nor there was necrosis. (Fig. 2) Immunohistochemical staining with neuroendocrine markers- chromogranin & synaptophysin showed positivity in tumor cells. Ki-67 proliferative index was noted to be less than 2 %.

A diagnosis of Carcinoid tumor of the appendix- Grade 1 was made with a TNM staging of pT2NxMx. After the incidental diagnosis of carcinoid tumor, post operatively, a CT abdomen & 24 h urine 5-hydroxyindole acetic acid (5-HIAA) level was assayed and found to be normal ruling out metastatic disease. Postop course was uneventfull and patient was discharged on the 5th day. Patient has been on follow up for 6 months and is disease free.

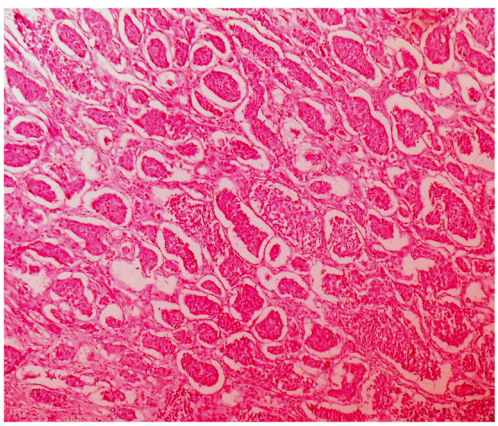

Fig. 2 Microphotograph showing nests of tumor cells with clear retraction spaces around them

Discussion

First recognized in 1867, Carcinoid tumors are rare malignant neuro-endocrine tumors. The reported incidence of appendiceal carcinoids in resected specimens in children in several studies range from 0.08 to 0.7 % [4–6].

Seventy-five percent of carcinoid tumors are localized to the apex, 20 % to the body and 5 % to the base of appendix and the median diameter of the tumor is 6 mm [7]. In the present case, the tumor was localized to the apex with a diameter of 13 mm.

Mean age at presentation of appendiceal carcinoid in children is 12–13 years, the youngest child reported is 3 years old. Female preponderance is seen in adults, probably as a result of increased abdominal procedures, however the incidence is not significantly higher in young girls compared with boys of same group [4].

Clinical presentation of appendiceal carcinoids is similar to that of acute appendicitis, but in some cases it is incidentally detected during surgery performed for another diagnosis or problem [2]. Carcinoid tumors located at the tip of appendix and measuring more than 10 mm usually present as acute appendicitis clinically, while tumors measuring more than 20 mm & located at the base present with signs of peritonitis clinically [2].

Carcinoid syndrome, which is commonly reported in adults has never been reported in any paediatric patient [8]. The symptoms are flushing, diarrhea, cardiac disease and are associated with liver and retroperitoneal metastasis which results in increased vasoactive substances such as serotonin, 5-hydroxytryptophan, bradykinin & histamine as they escape the first pass metabolism in the liver [8, 9].

Microscopically appendiceal carcinoid tumors are classified into 1. Classical appendiceal carcinoid tumors and 2. Goblet cell carcinoids. Classical appendiceal carcinoid tumors are well circumscribed, have solid nests of small, monotonous cells with scant cytoplasm, round nucleus with fine chromatin. Rarely the tumor may also show small acini with mucin rosettes, clear cells or vacuolated cells [10]. Retraction artifacts as seen in the present case are common and this finding should not be mistaken for lymphatic invasion.

Goblet cell carcinoids have a mixed phenotype with partial neuroendocrine differentiation and intestinal goblet cell morphology which stain positive with mucin stains. Goblet cell carcinoids bear worse prognosis and often require a right hemicolectomy [10].

The WHO 2010 classification subdivided appendiceal carcinoids into G1 and G2 based on the Ki-67 proliferative index and mitotic rate, both considered as predictors of metastasis and recurrence. Ki-67 of <2 % and mitotic rate of <2/10Hpf were classified as G1. Ki-67 of 3–20 % and mitotic rate of 2–20/Hpf were classified as G2 [11]. The present case belonged to grade 1.

The prognostic parameters are tumor size, proliferation rate, infiltration of the appendiceal wall and status of the surgical margins [10]. Prognosis of appendiceal carcinoid in children is good because tumors in this agegroup are small, less aggressive, rarely metastasize and usually present with symptoms of acute/chronic abdominal pain warranting early appendicectomy [7, 9].

Any incidental detection of carcinoid tumor after appendicectomy requires a general evaluation, measurement of serum levels of serotonin, chromogranin, urine levels of 5-hydroxynindole acetic acid (5-HIAA) and chest X-ray, USG abdomen/CT to ruleout lymphnode or distant metastasis.

Management of appendiceal carcinoid tumors depends on the tumor size and site. Small tumors (<2cms), located at the tip or mid appendix are treated by simple appendicectomy [7, 12]. No further surgical intervention is indicated if followup with 5-HIAA levels and abdominal CT is normal [7]. Extensive surgery, like a right hemicolectomy is recommended for large tumors (>2cms), tumors located at the base of appendix with positive margins, when the caecum is involved and for goblet cell carcinoids [2].

Incidental diagnosis of appendiceal carcinoids in children (<2 cms) and with no associated risk factors should still have regular annual clinical followup to monitor the outcome & prognosis [13].

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 case is an illustration to the fact that all appendicectomy specimens should be subjected to a routine histological examination.

Compliance with Ethical Standards

Source of Funding None

Conflict of Interest I, the corresponding author, on the behalf of all the authors, declare that we don't have any potential conflicts of interest, acknowledgements or source of funding to disclose and we have complied with the ethical standards in the present case.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards

Written Informed Consent Informed consent was obtained from all individual participants included in the study

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