Case Report

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Carcinoid tumour of appendix in a child: A rare case at an uncommon site

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SUMMARY

Carcinoid tumours of the appendix are uncommon incidentally detected tumours during histopathological examination following appendicectomy for acute appendicitis. Even though considered rare in children, they are the most frequently encountered tumours of the gastrointestinal tract. To our knowledge, carcinoid tumour of appendix in childhood has not yet been reported from Indian Subcontinent. The clinical presentation is similar to acute appendicitis and the signs and symptoms of carcinoid syndrome have not been reported in children. The prognosis of carcinoid tumour of appendix is excellent in children as the tumour is generally small in size and less aggressive with no metastasis. Simple appendicectomy is curative in most of the patients and long term follow up is debatable. We present here a case of carcinoid tumour of the body of appendix, which is an uncommon location in a 6-year-old child.

Key words: Body of appendix, carcinoid, Indian subcontinent

INTRODUCTION

Carcinoid tumours are rare neuroendocrine neoplasms, occurring with higher frequency in the appendix. They account for 32-57% of all the appendiceal neoplasms.[1] They are less common in children, with a reported incidence of 0.08%.[2] Appendiceal carcinoid tumours in children lack specific clinical features and their clinical presentation is similar to acute appendicitis. They are often diagnosed incidentally during surgery for appendicitis or during other abdominal procedures such as cholecystectomy, colectomy or salphingectomy in adults. However, more commonly, the diagnosis is made only after histopathological examination of the resected appendix. They usually behave as benign tumours although some of them have a potential for malignancy and can metastasize.[3] Appendicectomy alone is sufficient in most of the cases; however, extended surgery in the form of right hemicolectomy may be necessary in some. We report here a case of carcinoid tumour of the body of appendix in a young child which was diagnosed by histopathological examination.

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CASE REPORT

A 6-year-old girl child presented with pain in the right iliac region with fever, vomiting and malaise from 2 days. Physical examination and ultrasound abdomen favoured acute appendicitis. The patient underwent emergency appendicectomy. The resected specimen was 8 cm in length. The external surface showed a pale yellow bulbous swelling in the region of the body. Cut section revealed a solid yellow mass measuring 1.7 × 0.6 cm, located in the body of the appendix [Figure 1].

Microscopic examination showed features of acute appendicitis with the tumour in the mucosa, submucosa and muscularis propria. The tumour consisted of solid nests, cords and islands of uniform population of round to oval cells with eosinophilic cytoplasm, with fine nuclear chromatin [Figures 2 and 3]. Proximal surgical margin, mesoappendix and the resected mesenteric lymph node were free of tumour. Immunohistochemical staining with chromogranin [Figure 3 inset] and synaptophysin confirmed the neuroendocrine nature of the tumour. As per 7th edition of Tumor-Node-Metastasis (TNM) staging for neuroendocrine tumours of appendix,[4] the present case was under the category of pT1, pN0, pM0. Serum levels of serotonin and chromogranin, 24 hour urinary



Figure 1: Gross: Cut section of appendix showing a solid yellow mass measuring 1.7 × 0.6 cm in the body

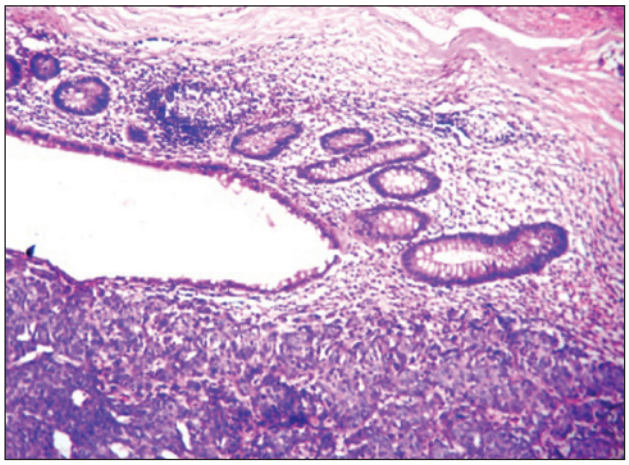


Figure 2: Photomicroscopy showing solid nests, cords and islands of tumour cells in mucosa and submucosa. (Hematoxylin and eosin stain −5×)

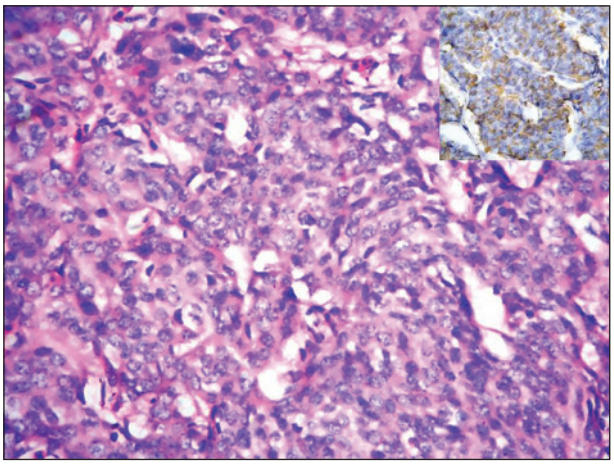


Figure 3: Photomicroscopy showing islands of uniform population of round to oval cells with eosinophilic cytoplasm, with fine nuclear chromatin. (Hematoxylin and eosin stain −40×). Inset – Chromogranin positivity in the cytoplasm of tumour cells (40×)

levels of 5-hydroxy indole aceticacid (5-HIAA) done post operatively, were found to be within normal limits.

DISCUSSION

Carcinoid tumours were first described in the ileum by a Swiss pathologist Theodor Langhans in 1867. Appendiceal location was described by Beger in 1882 and the term was coined by a pathologist, Siegfried Oberndorfer in 1907.[2]

Carcinoid tumours of appendix occur most commonly between 3rd and 4th decade. The frequency of carcinoid tumours is less in children, ranging from 0.2% to 0.5% of resected surgical specimens.[5] Dall'Igna *et al*., reported a frequency of 2 to 5 cases of carcinoid tumour per 1,000 appendicectomies.[2] A single large study conducted by Parkes *et al*., over a period of 30 years, showed an incidence rate of 1.14 per 1 million children per year.[3] However, the incidence and frequency between any two studies cannot be compared reliably as the decision for appendicectomy varies significantly from region to region and also from time to time.

Out of 800 appendicectomies done at our institute, only one case of carcinoid tumour of appendix was detected resulting in the frequency of 0.125%. Hager J, Parkes SE and Doede TH reported an incidence of 0.16, 0.085 and 0.169, respectively.[6]

The mean age of appendiceal carcinoid in children is 12-13 years,[5] with the incidence in younger children being even rarer; the youngest child reported is 3 years old.[6] Female preponderance is seen among adults probably as a result of increased abdominal procedures such as salphingectomy and colecystectomy. However, the incidence is not significantly high in young girls compared with the boys of the same age group, according to the study by Doede.[6]

Clinically, it presents as acute appendicitis and sometimes as chronic abdominal pain-the association is often coincidental.[5,7] Almost 70-90% of tumours are discovered incidentally during histopathological examination of resected appendix by pathologist,[8] as in the present case too. Carcinoids located at the tip, measuring <10 mm generally present with features of acute appendicitis, while those located at base, especially when large (>20 mm) may present with clinical symptoms of peritonitis.[9] Most frequent site of occurrence is the tip of appendix (75%), followed by body (20%) and base (5%).[10]

These tumours are classically well circumscribed, with solid nests of small monotonous cells with scant to moderate cytoplasm, round nucleus with fine chromatin.

Rarely, the tumour may also exhibit small acini containing traces of mucin, rosette formation, clear cells and vacuolated cells. Mitosis is exceedingly rare.[10] In the present case, the carcinoid tumour was located in the middle of the appendix with classical microscopic features. The unusual feature in the present case was the presence of extracellular pool of mucin in the muscularis propria, adjacent to the tumour. However, there were no viable mucin secreting cells, which are one of the predictive factors for aggressive behaviour and may require additional extensive surgery in the form of hemicolectomy. Additional sampling was done and mucinous cells were looked for to rule out the presence of any associated neoplasm. The mere presence of extracellular pool of mucin, situated adjacent to the tumour in the present case may be due to inspisated mucin secreted by the epithelial cells. The cell clusters are often surrounded by retraction artifact, giving a false impression of lymphatic invasion[10] and this phenomenon was observed in the present case also.

Prognosis of appendiceal carcinoid is good, because generally they behave like benign tumours, slowly growing, with symptoms of acute/chronic abdominal pain, resulting in early appendicectomy, and smaller tumours measuring <1 cm do not metastasize.[7] Tumour diameter is the most important parameter for predicting malignant potential; most carcinoids of tumour size <1 cm do not metastasize and appendicectomy is the best treatment.[7] For those measuring more than 2 cm, right hemicolectomy is indicated. For tumours measuring 1-2 cm in diameter, need for ileocaecal resection/right hemicolectomy is controversial, because frequency of metastasis is unknown. Conversely, ileocaecal resection or right hemicolectomy is suggested for tumours located at the base of appendix or intermediate type of tumours with production of mucin. Hence, assessment for surgery depends on site and size of tumour. Risk adapted follow up is suggested for patients at risk. Carcinoid with synchronous or metachronous, non-carcinoid malignant neoplasms have been reported. Also, patients with adenocarcinoma colon have been reported after long term follow up. This emphasizes the importance of careful search for presence of mucin producing cells at histopathology. Yearly follow up with detection of serotonin levels, and abdominal ultrasound is required in patients with tumour diameter measuring more than 5 mm diameter.[7] Further, computed tomography (CT) abdomen, chest, bone scan, serum serotonin and chromogranin levels, urinary 5HIAA levels are estimated to detect early metastasis or recurrence.

For tumours smaller than 2 cm and penetrating the serosa, treatment of choice is still appendicectomy whatever the location.[5] Metastasis of appendiceal carcinoid is very rare in children as most of the tumours are small in size and less aggressive.[9] In the present case too, the tumour measured 1.7 cm in diameter, but did not exhibit any metastasis.

Carcinoid syndrome is observed when retroperitoneal or liver metastasis co-exist which is rare and never seen in children. Urinary 5 HIAA levels is a good marker of endocrine activity of carcinoid tumours and high levels are found in patients with metastasis.[7]

After the diagnosis of carcinoid tumour in a resected appendix, regular follow up of patients with estimation of hormonal activity of chromogranin and serotonin checked every three months in first year, next every 6 months and annually after six years to look for clinical symptoms of carcinoid syndrome is recommended.[6] However, long term follow up in children is still a debatable issue considering their small size and non-aggressive behaviour.

Carcinoid tumour of appendix remains an incidental diagnosis. The present case highlights the continuing need for histopathological examination of appendix after every appendicectomy even if the appendix appears normal at laparotomy.

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