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A Case of Microscopic Carcinoid Presenting as Acute Appenditics

KEYWORDS: Microscopic carcinoid, Appendix, Histopathology

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ABSTRACT *Carcinoid tumours are uncommon malignant neuroendocrine neoplasms found mainly in the bowel and lung. They are the most common tumours of the appendix. Commonly there are found incidentally on appendectomy and rarely reported in children. We reported a 13-year-old male who presented with the clinical picture of acute appendicitis. Carcinoid tumour was diagnosed on histological examination of the removed appendix. The purpose of reporting this case of carcinoid appendix is to bring the awareness of the diagnosis so that appendix after removal is submitted for histopathological examination even if the appendix looks normal grossly.*

1.Introduction:
Carcinoid tumours were first reported by Otto Lubarsch in 1888.[1] In 1907, S Oberndorfer introduced the term carcinoid tumour to describe a midgut tumour that was morphologically distinct and less aggressive in behaviour than intestinal adenocarcinoma. Carcinoid tumours are among the most puzzling tumours in clinical practice. They are neuroendocrine tumours derived chiefly from enterochromaffin cells. [2] They are commonly seen in the third to fifth decade. [3,4] The occurrence is estimated to be approximately 0.05 per 100, 000 although autopsy reports suggest that it may be as high as 2 per 100,000. [5] Clinical presentation of these tumours is alike to acute appendicitis, but usually they are found incidentally on appendectomy, surgery or histological examination of the surgical specimens.[6] 75% of the carcinoids occur in the gastrointestinal tract (GIT), 25% in bronchopulmonary system while the remaining 1% is reported in ovary, thymus, testis, liver, spleen. Appendicular carcinoids have the most favourable prognosis of all carcinoid tumours. [3.]

2. Case Report
A 13-years-old male patient presented with complaints of abdominal pain in right lower quadrant,nausea and decreased appetite for two days. Physical examination revealed a healthy looking male child with mild distress, a temperature of 100.8 degrees, and blood pressure 110/60 mmHg. He had severe direct and rebound tenderness in the right lower quadrant and had no hepatomegaly or splenomegaly or lymphadenopathy, and the rest of his physical examination was normal. Blood investigations were done .The TC was slightly high (13,000) with neutrophilia of 78%. The patient was operated for acute appendicitis. Grossly the appendix measured 8.5 X 0.7 X 0.7 cms. Outer surface showed congested blood vessels. Cut surface showed patent lumen with faecal matter. No tumour was found grossly. Microscopically, the sections from the tip of the appendix showed appendicular mucosa with one focus showing tumour made up of monotonous medium sized round to oval cells with round to oval nuclei with salt and pepper chromatin. The tumours were arranged in ribbon like cords. (Figure: 1) The tumour was seen infiltrating the submucosa and superficial part of the muscle layer. Immunohistochemistry for Neuron specific enolase was done and it came positive. (Figure: 2)

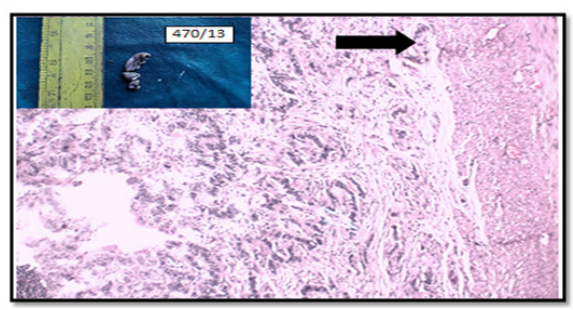

Figure 1 showing tumour cells are arranged in ribbon pattern (arrow) H& E X 40 Inset showing gross specimen of appendix

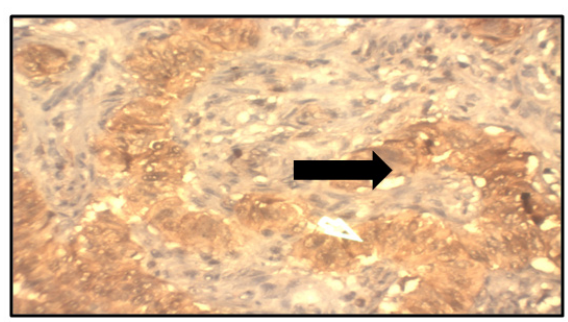

Figure :3 IHC 40 X Tumour cells showing diffuse positivity for NSE (arrow)

3. Discussion:
Carcinoid tumour was first recognized in 1867.[7] Carcinoid tumours account for 50-77% of all appendiceal neoplasms.[8, 9] Around 19% of all carcinoids are located in the appendix. The mean age at presentation of carcinoid is 32-43 years.[10] The present case was reported in a male child of 13 years old. Acute appendicitis is more common in pre-teen and teenage boys, while female to male occurrence in carcinoid of the appendix is 2:1.[11] In a series of patients younger than 15 years of age, 14 of 18 appendiceal carcinoid tumours were found in girls.[12]. Mostly carcinoid tumours in children arise in the appendix; but they can also occur in other primary sites including the small intestine, bronchus and others .[13] The clinical presentation underestimates the frequency, as many carcinoids remain asymptomatic.[14] But it can present as acute appendicitis clinically.[6] But it can present as acute appendicitis clinically as in the present case. Recurrent episodes of abdominal pain were reported in many cases which indicate

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partial obstruction of the appendiceal lumen by a tumour [15] In another two reported cases the patients presented with clinical signs of peritonitis without previous episodes of acute abdominal pain [16] Cases can present as flushing, diarrhoea, cardiac disease and usually associated with liver or retroperitoneal metastases. [17, 18] The tumour 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% .[13] In our case it was located in the tip. The median tumour diameter is 6 mm but in the present case grossly no tumour was found .Usually grossly carcinoid tumours appear as firm yellow colour after fixation and well circumscribed mass.[10]. Microscopically, the tumour cells were showing features of carcinoid tumour in one focus at the tip of the appendix. Microscopically, classic carcinoid tumours are formed by solid nests of small monotonous cells with occasional acinar or rosette formations.[19]. A minority of classic carcinoid tumours are composed of enteroendocrine cells rather than enterochromaffin cells. They tend to feature glandular structures rather than solid nests. The tumour cells were arranged in ribbon like cords in our study.[20] In our case immunohistochemistry which was done for NSE showed strong positivity and this helped in confirmation of the diagnosis.

In conclusion, carcinoid tumour is rare in children, even though it is a common tumour in appendix. Clinically it can mimic the features of acute appendicitis. Clinicians should be aware of carcinoid tumours of the appendix since early diagnosis of these tumours is important in preventing morbidity and mortality because it can be metastatic.

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