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Carcinoid of the appendix in a six year old child – A case report

Dr.S.Srikanth*, Assistant Professor , Dept Of Pathology, Prathima Institute Of Medical Sciences, Karimnagar, Telanagana State, India.

Abstract:
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. 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.
Keywords: Carcinoid ,appendix,child

Case report:
A 6 year old female patient came to emergency department with acute onset of pain in right lower abdomen. She had tenderness and rebound tenderness in right iliac fossa. She also complains of fever and 2 episodes of vomiting. Her total count was 15,000 cmm3. Her other blood investigations was normal. Ultrasound abdomen was consistent with our clinical diagnosis of acute appendicitis. Patient was taken up for emergency appendectomy. Intraoperatively acute inflamed appendix was present. Appendectomy was done. Histopathology report was acute appendicitis with carcinoid tumor involving the tip (<1 cm) and extending up to muscle layer. (Figure 1 &2) The proximal surgical margin is free from tumor.

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. [1]

Carcinoid tumours of appendix occur most commonly between 3 rd and 4 th decade. The frequency of carcinoid tumours is less in children, ranging from 0.2% to 0.5% of resected surgical specimens. [2] 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.

The mean age of appendiceal carcinoid in children is 12-13 years, [2] with the incidence in younger children being even rarer; the youngest child reported is 3 years old. [4] Female preponderance is seen among adults probably as a result of increased abdominal procedures such as salphingectomy and colecystectomy.

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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. [4]

Clinically, it presents as acute appendicitis and sometimes as chronic abdominal pain-the association is often coincidental. [2], Almost 70-90% of tumours are discovered incidentally during histopathological examination of resected appendix by pathologist, [5] 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. Most frequent site of occurrence is the tip of appendix (75%), followed by body (20%) and base (5%).

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. In the present case, the carcinoid tumour was located in the middle of the appendix with classical microscopic features.

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. 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. 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. 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. [2] Metastasis of appendiceal carcinoid is very rare in children as most of the tumours are small in size and less aggressive. In the present case too, the tumour measured 1.7 cm in diameter, but did not exhibit any metastasis.

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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.

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. [4] 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.

Figure legends:

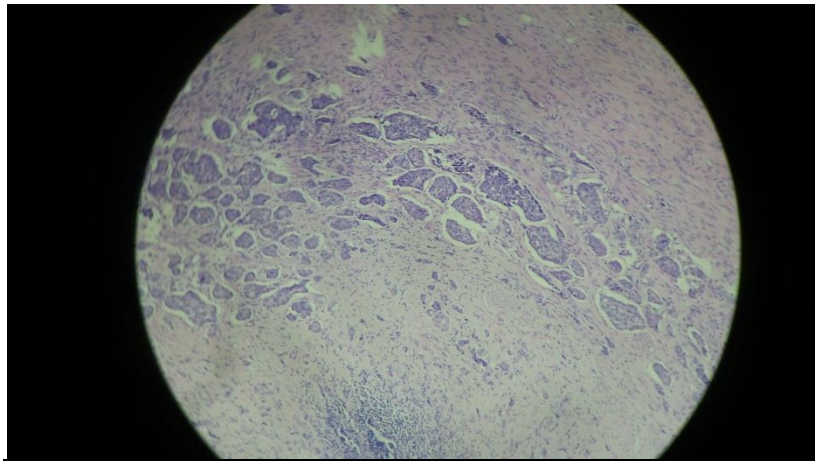

Figure 1: Section showing islands of uniform population of round to oval cells with moderate amount of eosinophilic cytoplasm with fine nuclear chromatin (H&E,x10)

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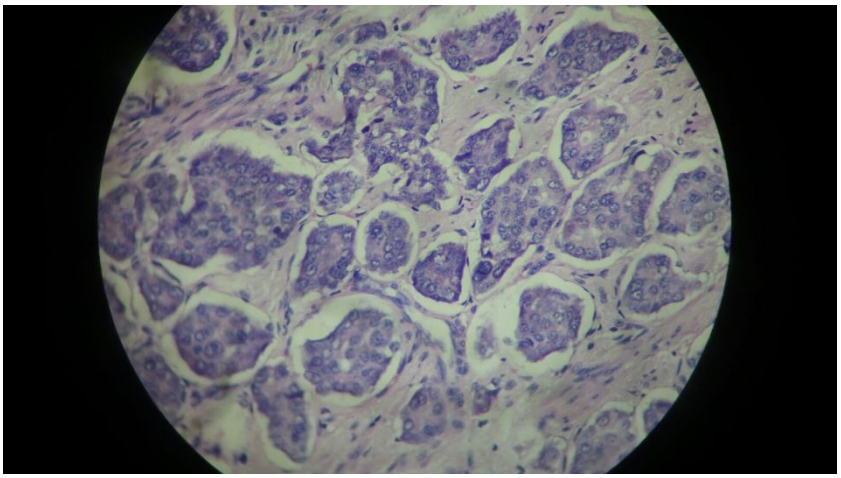

Figure 2: Section showing monotonous population of tumour cells.(H&E,x40)

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