ABSTRACT
Carcinoids are the most common tumors of the appendix. The prevalence of these tumors is more in children. Clinically appendiceal carcinoids present as acute appendicitis, although in many cases the tumor is diagnosed incidentally during an operation. The diagnosis is confirmed on histology. The prognosis in patients with local disease is excellent. In small lesions, ‘Isolated Appendicectomy’ is considered as the most appropriate treatment, while in larger lesions ‘Right Colectomy’ should be performed. We report a case of a carcinoid tumor appendix measuring 0.7 cm in diameter, in a 21 year old female, which was diagnosed on histology. Appendicectomy was done with a clinical diagnosis of ‘Acute Appendicitis’. Carcinoid tumor was incidentally diagnosed on histology. The patient is considered free of disease as the prognosis of tumor less than 2cm local is excellent.

KEY WORDS: appendix, carcinoid tumor, morbidity, mortality

INTRODUCTION:
The term “karzinoid” was first used by Oberndorfer in 1907 to a type of tumor which grew slowly and appeared to be more benign than adenocarcinomas. The description of the malignant carcinoid syndrome Cassidy’s report in 1934. The endocrine potential of the carcinoid tumor, and its subsequent relationship with the varied clinical manifestations, however, was recognized after the discovery of serotonin by Rapport et al. In 1948, and the isolation of serotonin from a carcinoid tumor by Lembeck in 1953. In 1954, Thorson and coworkers wrote about the endocrine properties and clinical manifestations of carcinoid syndrome like flushing, diarrhea, cardiac disease which have been rarely reported and usually associated with liver or retroperitoneal metastases[1]. In these cases an increased urinary excretion of 5-HIAA has been documented in monitoring disease and progression of tumors.

Case Report
INTRODUCTION:
Carcinoid tumors are the most common tumors of the appendix[2], accounting for more than half of all appendiceal malignancies and found in seven of every 1,000 appendectomy specimens, they account for 5% of GI carcinoids and are more common in women than men. The mean age at presentation is 49 years. This may reflect the common patient age at appendectomy, when the tumors are often incidentally discovered. Approximately two-thirds of appendiceal carcinoid tumors arise in the tip of the appendix, where they are unlikely to cause symptoms of obstruction. Ten percent of appendiceal carcinoids occur in the base where they are more prone to obstruct the appendix and cause acute appendicitis. It has been proposed that the incidence of appendiceal carcinoid tumors parallels the activity of subepithelial NE cells, the source of these tumors. The density of these NE cells peaks in the second decade of life and then decreases.[3]

CASE PRESENTATION:
A 21-years female patient presented with abdominal pain in right lower quadrant (RLQ) and nausea since last 30 days and fever off and on since 5
days. Physical examination revealed a healthy looking female with a mildly elevated temperature 38°C, blood pressure 112/60 mmHg and pulse rate 76/min. The patient showed signs suggestive of acute appendicitis, due to direct and rebound tenderness in the RLQ. Rectal examination revealed mild tenderness but no blood or palpable mass was observed. Hematocrit was 29.6%, white blood cells 5000/cmm with neutrophil prevalence 76%, platelets 446000/comm., erythrocytes sedimentation rate 55 mm/hr, CRP 1.44 mg/l, while the remaining blood analysis was normal. Abdominal ultrasound revealed appendiceal inflammation. Laproscopic appendisectomy was done and patient discharged on second post operative day. On gross examination appendix was found to be inflamed with mild distension of the tip. Cut section showed well circumscribed yellowish mass measuring 0.7 cm in diameter (Figure 1). No local or regional enlarged lymph nodes were found. The histological examination of H&E (Haematoxylin & eosin) stained section revealed a typical carcinoid tumor of the appendix and tumor-free margin in all the sections. The tumor consisted of small homogenous neoplastic cells that were mostly arranged in islet formation and infiltrated few areas of the muscular layer, at places up to the serosa. The mitotic activity was insignificant. There was a coexisting acute appendicitis and periappendicitis (Figure 2).

DISCUSSION:

Carcinoid tumors of the appendix are relatively uncommon neoplasms. Although considered rare pathology, these are the most frequent tumors of the gastrointestinal tract in childhood and adolescence. They are usually benign neoplasms and the uncommon occurrence of metastasis is related to the primary tumor size and depth.

The reported incidence of appendiceal carcinoids in several studies ranges from 0.08 to 0.7% in surgical specimens. Willox in 1964 suggested that 0.2–0.5% of surgically removed appendices in children contained CT. D’Aloe et al. estimated that carcinoid tumors of the appendix occur in 1:100,000 to 169:100,000 children. The clinical presentation of the appendiceal carcinoids is similar to that of acute appendicitis, but in some cases the disease is incidentally found during surgery performed for another diagnosis or problem. Recurrent episodes of abdominal pain reported in many cases may indicate partial obstruction of the appendiceal lumen by a tumor. In two reported cases the patients presented with only clinical signs of peritonitis without previous episodes of acute abdominal pain. The tumor is more common in females with a mean age of 12 to 13 years. Symptoms of the carcinoid syndrome like flushing, diarrhea, cardiac disease have been rarely reported and usually associated with liver or retroperitoneal metastases. In these cases an increased urinary excretion of 5-HIAA has been documented in monitoring disease and progression. Our patient had no symptoms related to carcinoid syndrome, neither metastatic spread nor 5-HIAA increased excretion. The majority of carcinoid tumors are discovered during the histological examination of the surgical specimen incidentally and rarely suspected before this examination.

At present, the site and the size rather than the depth, are used for the assessment of the tumour.
In 75% of cases the tumor is localized at the apex of the appendix; 20% and 5% affect the mid portion and the base respectively. The median diameter of the tumor is 0.7 cm. In our case the diameter of the tumor was 0.7 cm. Generally, carcinoid tumors located at the tip of the appendix and measuring less than 1 cm, usually mimick the clinical presentation of acute appendicitis, while tumors measuring more than 2 cm and located at the base of the appendix may present with clinical signs of peritonitis. The prognosis is directly related to the tumor size. If the tumor is smaller than 2 cm and has perforated the serosa, the treatment of choice is appendectomy, whatever the location. Other reports suggest that neoplasms with these characteristics do not tend to relapse. Tumors measuring 2 cm or more in diameter may have widespread metastases upon detection. The invasive properties of these tumors are well-known, but the presence of lymph node metastases is reported in only 4% to 5% of paediatric cases. The prognosis is excellent. It should be mentioned that carcinoid is not infrequently associated with MEN1 and loss of 11q, sometimes independently of the MEN1 gene (11q13), suggesting loss of MEN1 or another tumor suppressor gene is responsible for the condition. However, this is usually isolated to foregut carcinoids.

Metastasis of an appendiceal carcinoid tumor is very rare in children probably because most reported tumors in this age group are small and less aggressive. In this case the tumor involved the layers of the wall and extended up to the serosa without distant metastasis.

The treatment of carcinoid tumors in the appendix depends on the size and the site of the tumor. Tumors smaller than 2 cm can be adequately treated by appendectomy, while right hemicolectomy is recommended for pediatric patients with appendiceal carcinoid tumors larger than 2 cm, especially when the mesoappendix is involved or in cases with residual tumors at the margin of resection. Our patient had an appendiceal carcinoid tumor of 0.7 cm in size, with tumor free-margin in all sections; in such patients simple appendectomy is considered the adequate treatment.

CONCLUSION:

We reported case of an appendiceal carcinoid tumor with asymptomatic i.e. free of carcinoid syndrome in a 21 years female with the diameter of 0.7 cm which was treated with isolated appendectomy and the patient is disease free after appendectomy as the prognosis of tumor less than 2 cm local is excellent. Carcinoids tumors constitute an important group of neoplasms’s demanding careful assessment and discerning management to decrease morbidity and mortality.

REFERENCES:


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