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### RESEARCH ARTICLE

#### CARCINOID TUMOR OF THE APPENDIX, MANAGEMENT AND PROGNOSIS A CASE REPORT.

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#### Abstract

Carcinoid tumors of the appendix are usually asymptomatic. Hence, typically, it is just an incidentally encountered pathology following an appendectomy. However, carcinoid tumors are quite rare incidents in clinical practice (1, 2).

Although it can affect any age, this type of neoplasm presents in adolescents more than any other age group (1). Here we report the case of a 15-year-old girl who presented with clinical features suggestive of acute appendicitis. Laparoscopic appendectomy was carried-out. Subsequently, the patient recovered uneventfully. Nevertheless, the histopathologic examination revealed findings consistent with an appendicular carcinoid tumor. In this report, Presentation of this neoplasm, its diagnosis, management, and prognosis were reviewed.

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#### Introduction:-

Carcinoid tumors are rare, slow-growing, neuroendocrine neoplasms (3). These tumors are supposed to have a neuroectodermal origin and thus classified as part of the other amine precursor uptake and decarboxylation (APUD) neoplasms(2). Traditionally, it is divided into foregut, midgut, and hindgut tumors(4). The appendix is the most frequent incidence site, followed by the rectum, ileum, lungs, bronchi, and stomach(3). Notwithstanding that, cases were reported in other body parts like gallbladder and even testis(5, 6).

However, carcinoid tumor of the appendix, unlike small bowel carcinoid, is often silent, and rarely shows any specific clinical presentation. It is predominantly an incidental finding during appendectomy for appendicitis or some other surgical procedures (1-3). Diagnosis of appendiceal carcinoid tumor is a real clinical challenge (3). It is a good suggestion that during any laparotomy, the appendix has to be palpated carefully, looking for a firm, bulbar mass in order not to miss the presence of a carcinoid tumor (2).

Appendiceal carcinoid tumor incidence is equal in both sexes with a slight female preponderance. It can occur at any age. Starts from early childhood, peaks in young adults then steadily decreases in patients older than 60 years. The

latter trend parallels the frequency of the performed appendectomies (1), as the same justification behind the slightly higher female predominance(7).

In foregut tumors, deletions and mutations of the MEN1 gene on chromosome 11 are common, whereas midgut carcinoids show genetic changes on chromosome 18. On the other hand, hindgut tumors, in general, are associated with a low transforming growth factor- $\alpha$ . Additionally, chromogranin A (Cg A) is a general tumor marker for all types of the carcinoid tumor (4). Nevertheless, diagnostic procedures are unlikely to help in diagnosing the usually small size appendiceal carcinoid. And it is only helpful in the patient follow-up(8).

#### **Case report:-**

A 15 years old female presented with anorexia, vomiting, and central abdominal pain of acute onset. Gradually, the pain shifted to the right iliac fossa. However, it was not associated with bowel habit change nor fever.

She had no significant personal medical or surgical history. However, her mother had appendectomy a year before, but with histopathological findings suggestive of a simple acutely inflamed appendix.

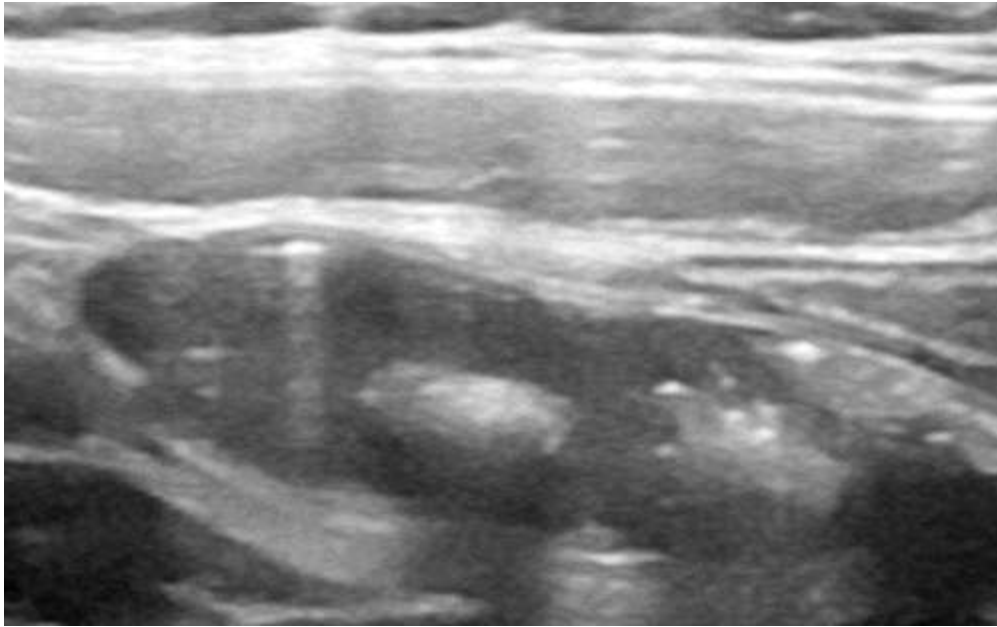
Clinical examination revealed mild localized tenderness and rebound tenderness in the right iliac fossa, with no other significant sign. Nevertheless, inflammatory markers and urine analysis were normal. The calculated Alvarado score was 5 points.

Subsequently, imaging study was supportive of the clinical diagnosis. As abdomen ultrasound showed a distended incompressible appendix lumen measuring 11 mm in diameter, contains multiple appendicolithses, and associated with a surrounding thin rim of intraperitoneal fluid, reactive mesenteric lymph nodes and probe tenderness.

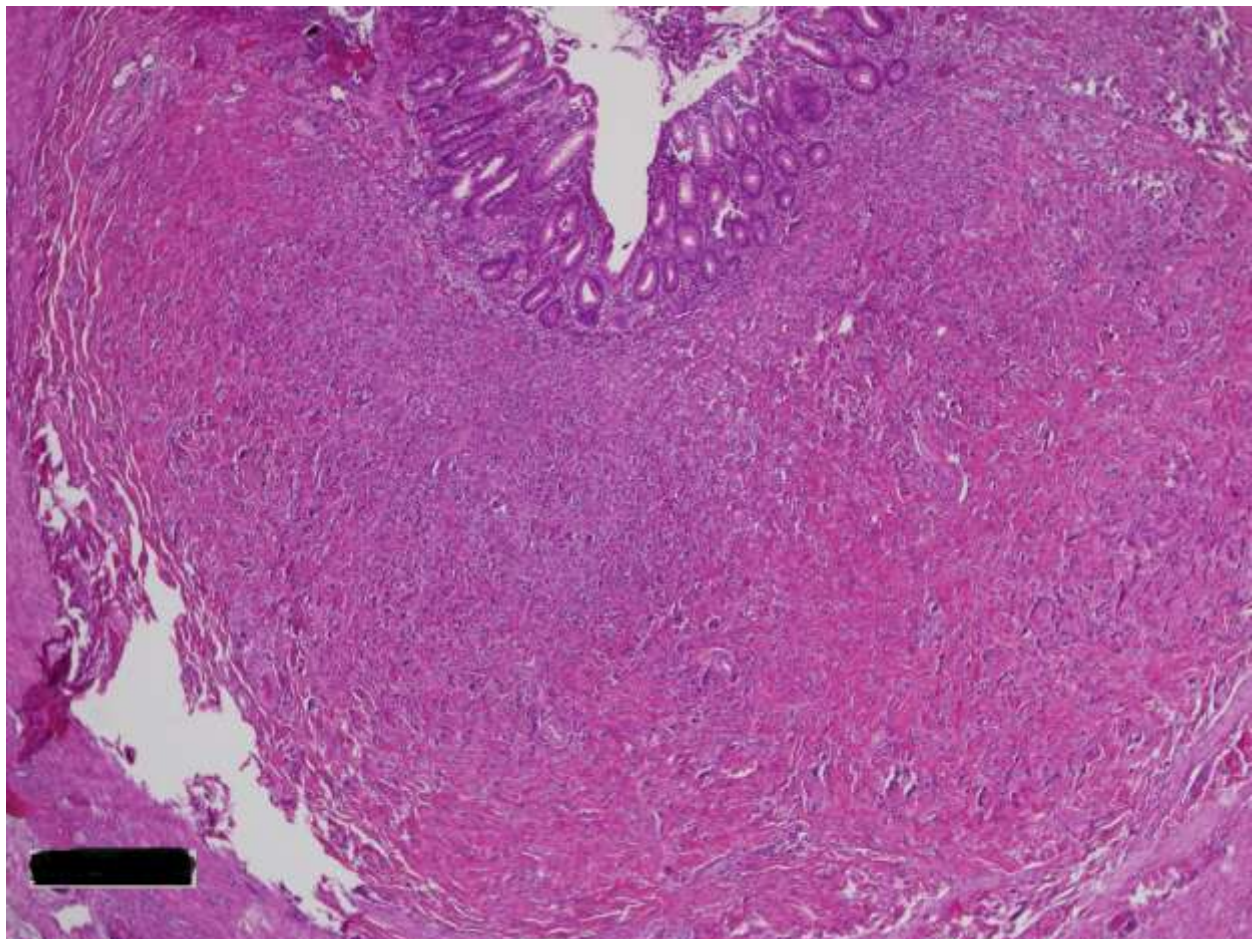
Laparoscopic appendectomy was carried-out. The patient's recovery was uneventful. Subsequently, she was discharged home in a good condition.

The specimen was sent for histopathological examination which was reported as acute appendicitis with associated classic type (grade 1) carcinoid tumor of the appendix. Tumor size was 4 x 4 mm located in the mid-portion of the appendix, confined to the submucosa and inner layer of muscularis propria. However, serosa was uninvolved. Tip, base, and mesoappendix were free of tumor. The Non-neoplastic appendix showed acute inflammation. Immunohistochemistry markers like Synaptophysin, Chromogranin, and CD56 were positive in the tumor cells. And Ki67 showed low proliferative index (1%).

Consequently, Oncology consultation was sought, and work-up was conducted. Contrast CT scan for the chest, abdomen, and pelvis, along with CEA and CA 19-90 were all normal. So far, during follow up in the oncology clinic, the patient is doing perfect with no related complaint.

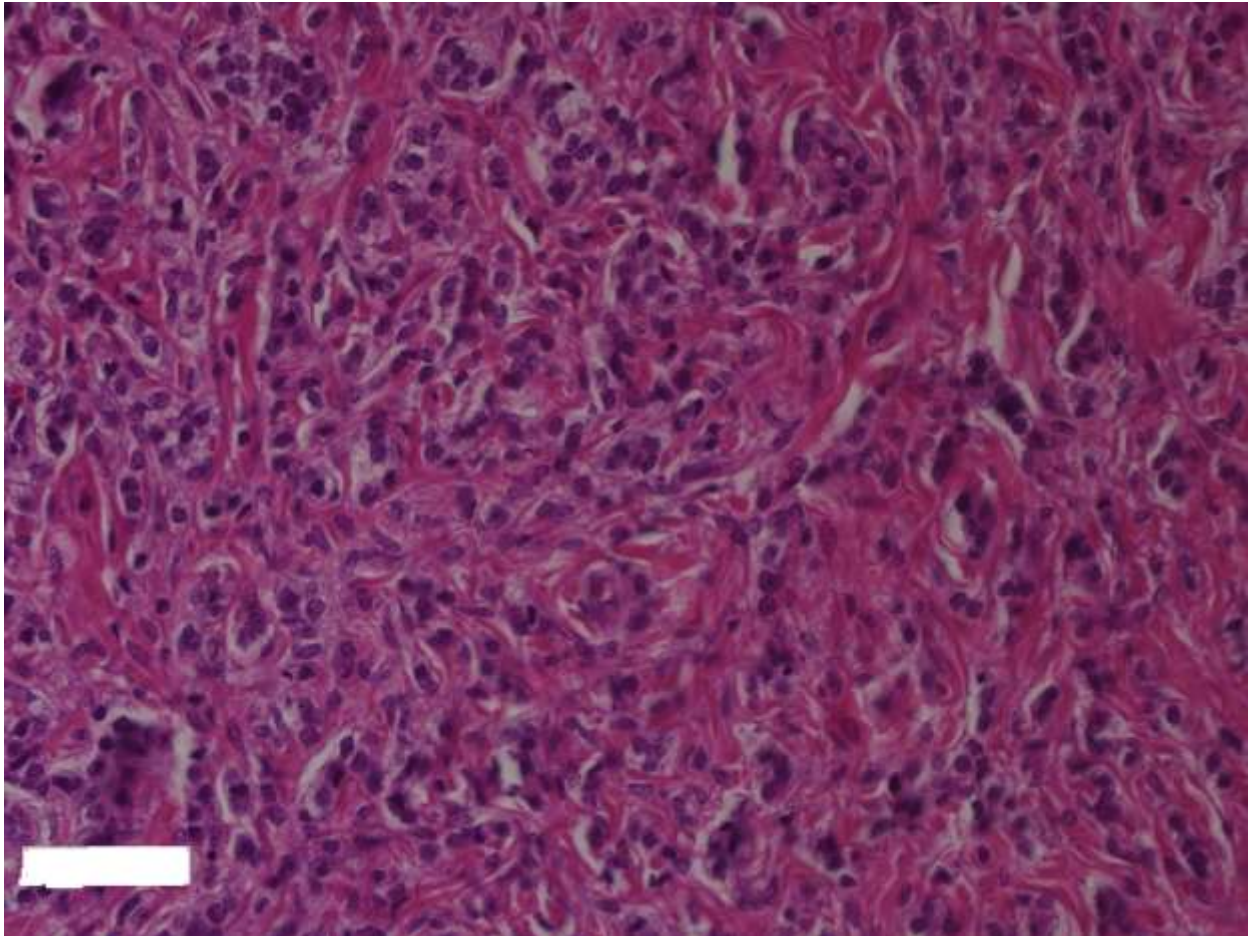


USS view showing a distended appendix with appendicoliths in the lumen



LPF microscopic view showing the mucosa and the carcinoid tumor cells





HPF microscopic view showing the carcinoid tumor cells



Microscopic view showing the positive Chromogranin test on the tumor cells

### Discussion:-

Carcinoid tumor of the appendix is usually an incidental finding. And it doesn't express any specific clinical sign. As mentioned earlier, the diagnosis is based on the histopathology report, and so the prognosis prediction.

The reported case presented with a typical clinical picture of appendicitis. There wasn't any specific sign of carcinoid tumor. Therefore, simple laparoscopic appendectomy was carried out. Carcinoid tumor discovered incidentally. Subsequently, full screening was performed and treatment plan was decided accordingly.

Tumor size and extent of invasion are the most important prognostic factors. It is unlikely to have metastasis with the tumors  $\leq 2$  cm in their maximal diameter (1, 2, 8). Also, the microscopic lymphatic invasion and the superficial (but not the deep) serosal or mesoappendiceal invasion don't carry the risk of recurrence after appendectomy (1, 2, 8). Similarly, there is no data indicating that location at the mid-third or at the base of the appendix carry a negative prognostic effect, although (70%) of the tumors are located at the tip of the appendix(8).

In contrast, the tumor greater than 2 cm, deep ( $>3$ mm ) serosal or mesoappendiceal invasion, or involvement of resection margin, all possess a relevant risk of recurrence and further surgical procedures are warranted (8).

Longtime, there was agreement-like that appendectomy is a sufficient treatment for all tumors sizing  $\leq 1$ cm and R0 resection. While some controversy was there regarding the tumors of 1-2 cm in size (2, 9).

Accordingly, an appendectomy can be considered as a complete treatment for this reported case; a small size (4 X 4mm) tumor, located at the middle part and no invasion in the serosa with free mesoappendix.

Currently, most of the guidelines consider carcinoid tumor cured after appendectomy with R0 resection, for the well-differentiated tumors with diameter  $\leq 2$  cm (1,9). The same also was suggested by the Consensus Conference of the European Neuroendocrine Tumor Society (ENTS). Hence, no follow up is required regardless of the location of the tumor (8). However, in tumors greater than 2 cm, positive or unclear resection margins, and deep ( $>3$ mm) mesoappendiceal or angioinvasion, right hemicolectomy is the advocated treatment (1, 8, 9). Moreover, CT of the abdomen and somatostatin receptor scintigraphy may be performed. On the other hand, factors like; high proliferation marker, vascular involvement, and deep mesoappendiceal infiltration are believed to argue for follow-up investigations (8).

Follow-up for patients considered cured is not justified and only one CgA determination 6–12 months postoperatively has been suggested. However, other patients should be investigated 6 and 12 months postoperatively and then yearly lifelong (8).

So this patient was referred for follow up at the oncology department.

### Conclusion:-

Carcinoid appendix is a rare, silent neoplasm with a wide-range of incidence-age extending from early childhood to over 60 years old, with the peak incidence at adolescence.

It is mostly diagnosed incidentally. Hence the need for Palpating the appendix during every Appendectomy, looking for the presence of any firm bulbar nodule, or, at least to feel the removed appendix before sending to the laboratory in case of laparoscopy.

Furthermore, the histologic findings are crucial to diagnose, to tailor the management plan and to decide if further hemicolectomy or follow up are needed.

Appendectomy is the satisfactory treatment for the majority of cases. While CgA 6-12 months after the procedure is the only recommended follow up investigation.

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