

# Know the Behavior of Appendiceal Carcinoid

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## Abstract :

Appendectomy is a most common emergency operation in general surgical field. The most frequent underlying pathology is acute appendicitis, however other pathologies are involved at times. Carcinoid as a neuroendocrine tumor is one example of such pathologies that need further evaluation and treatment. The recent years have seen the incidence of carcinoid tumor of appendix grow notably. The awareness of detailed behavior of appendiceal neuroendocrine tumor and their biologic nature could lead to treatment options of adequate validity<sup>1</sup>. Here we are presenting a case of recurrent appendicitis, who's histopathological report revealed carcinoid of appendix.

**Keywords :** Carcinoid appendix, behavior

## Case Report :

An 18yr old female patient presented to surgical OPD with dull aching right lower quadrant abdominal pain for the past one & half month, which was on and off. On examination she was afebrile with stable vitals. Abdominal examination revealed mild tenderness in right iliac fossa. Routine blood investigations were within normal limits. Sonography of the abdomen revealed thick appendix (12mm diameter), no abdominal or pelvic collection, no abdominal lymphadenopathy. With a probable diagnosis of recurrent appendicitis, appendectomy was done under spinal anesthesia. Grossly

Follow up at 3 months showed normal abdominal scan and normal 24hr urinary 5-HIAA.

## Discussion :

Carcinoid tumor originating from neuroendocrine cells can be enteric or non-enteric origin. "Kerzinoïd" is a word was first used by Obendorfer in 1907 to explain a tumor behaving in a fashion that is more benign than malignant<sup>4</sup>. In 1975 Goldwin reported on a wide range of sites for carcinoids including lung, ovary, biliary system and whole of GI tract.<sup>5</sup>



Fig 1: Gross appearance

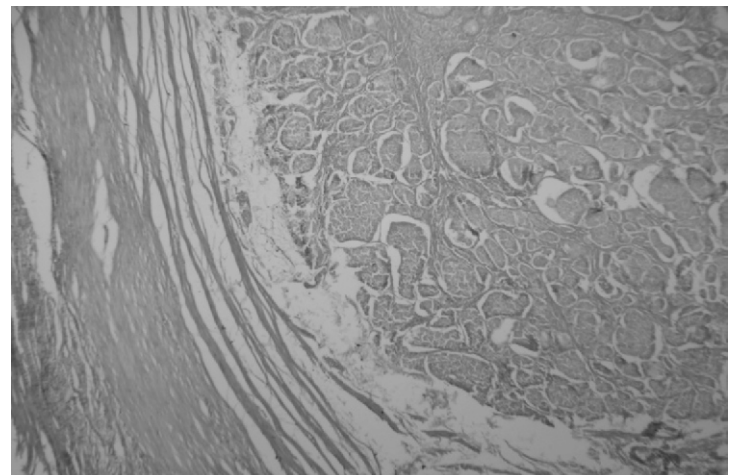


Fig 2 : H&E LP -

Uniform cells in groups with pepper like chromatin

appendix was thick, fibrosed and yellowish-white and was sent for histopathological study. Post-operative period was uneventful. Histopathology revealed presence of carcinoid measuring 1 cm at the tip of the appendix with proximal surgical margin free from tumor.

**Carcinoid Syndrome** consists of a group of symptoms and findings on physical and laboratory examination that are sometimes caused by the potent hormones produced by carcinoid tumors. They are widely distributed in the body but found in greatest amounts in the small intestine and then in decreasing frequency in the appendix, rectum, lung, pancreas and very rarely in the ovaries, testes, liver, bile ducts and other locations. Of all carcinoids, those arising in the appendix are the most benign, having only

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very rare distant spread. A carcinoid is found usually by accident in 1 of every 200-300 appendices removed at surgery.

The type of symptoms will depend on where the tumor started, where it may have spread to, and whether it produces a hormone known as serotonin. Non-functioning carcinoid tumors are so slow growing that many years may pass between the onset of any symptoms and the diagnosis, and produce vague symptoms. Biochemical markers like 5-HIAA in 24hr urine [88% sensitive, 35% specific], Plasma chromogranin A [very sensitive, less specific, Gold Standard], Imaging like Somatostatin Scintigraphy with <sup>111</sup>Indium-octreotide [90% sensitive] are helpful in diagnosing carcinoid.

Carcinoid tumors vary greatly in their size, location, symptoms, growth and spread. Therefore the treatment in each case should be individualized to what is best for each particular patient. With complete removal of all of the tumor tissue, is the first and best treatment when it is possible, and if detected early can result in a complete and permanent cure<sup>6</sup>. If it is possible to remove the tumour completely no other treatment may be necessary. Surgery may still be possible for metastatic tumors (spread to only one or two areas) because of the slow growth of the tumour.

Chemotherapeutic drugs used alone have been disappointing but a number of combinations of these drugs have been beneficial. Radiotherapy is usually given to treat symptoms, such as pain, which may occur if the tumour has spread to the bones. Somatostatin analogues [Octreotide, Lanreotide] work by reducing the production of hormones by the tumour, and can help to reduce the flushing and diarrhoea. But are now believed to sometimes inhibit or even reverse growth of the tumors. This has become the mainstay of treatment for most carcinoid tumors, with or without the Carcinoid Syndrome.

Interferon-Alfa is sometimes helpful in shrinking or slowing the growth of metastatic neuroendocrine cancers and improving symptoms of carcinoid syndrome. Its usefulness is sometimes limited by its flu-like side effects. Advances in diagnostic methods and surgical techniques have allowed more active management and improved prognosis. The prognosis for patient with completely resected localized disease is excellent. Factors that determine the clinical course and outcome of patients with GI carcinoid tumors are complex and multifaceted and include the site of origin, the size of the primary tumor, and the anatomical extent of disease. In general, patients with carcinoid tumors of the appendix and rectum experience longer survival than patients with tumors arising from the stomach, small intestine, and colon.

### Conclusion

Appendiceal carcinoid tumor occurs most often as acute appendicitis. In most cases, it is found incidentally during appendectomies and its diagnosis is rarely suspected before histological examination. A Careful evaluation of diagnosis is further needed for treatment indications. Extended surgery or further medical treatment must be planned for patients with defined risk factors. Even though the appendiceal carcinoid has a favourable prognosis, the synchronous or metachronous colorectal cancers can occur. Hence, surgeons have to be aware of the possibility of worst case progression of disease.

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