

Neuroendocrine Tumor of Appendix-A Case Report

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Abstract

Neuroendocrine tumors (NETs) are characterized by their ability to secrete hormones. NETs can develop in any organ of the gastrointestinal system. NETs of appendix are rare in occurrence with an estimated incidence of 0.15 to 0.6 cases/year per 100,000 people and are often seen in young adults with a higher male preponderance. NETs of the appendix are usually asymptomatic and are discovered incidentally after appendectomy. Treatment is by surgical removal of the tumor with a healthy margin in accordance with the principles of oncology.

Here we report on a case of a 40-year-old male patient. Admitted with sudden onset right iliac fossa pain with no fever. Emergency abdominal ultrasound revealed a 1-cm dilated non-peristaltic blind loop of bowel with moderate fat stranding suggestive of acute appendicitis. The biochemical work-up revealed leukocytosis with a predominance of neutrophils. The patient underwent laparoscopic appendectomy. Histopathological examination showed an incidental neuroendocrine tumor of appendix Grade 1, less than 2 cm with no lympho-vascular invasion with a free base.

Keywords: appendix; carcinoid tumor; neuroendocrine tumor; right hemicolectomy

Introduction

Neuroendocrine tumors (NETs) are mainly arise from enterochromaffin cells present in the gastrointestinal tract and the respiratory system. Neuroendocrine tumors of the appendix are the third most common GI neuroendocrine tumor after small intestine and rectum. Appendicular neoplasms are rare in occurrence. Their presentation is mostly asymptomatic. Tumors of the appendix are mostly incidental and are identified in patients operated on for acute appendicitis. They are detected in about 1% of appendectomy specimens on histopathological examination [1]. Current guidelines suggest that a simple appendectomy is an adequate treatment for NETs if they are less than 1 cm in size, and a right hemicolectomy is recommended for those larger than 2 cm involving the base and with higher grades. However, the ideal treatment for tumors between 1 and 2 cm is still unclear.

Case Report

Our case was a 40-year-old gentleman who presented sudden onset right lower abdominal pain since few hours with no history of fever, reduced appetite, nausea or vomiting. Pain was localized to

the right lower quadrant and was non-migratory. Patient did not complain of any increased frequency of urination or burning sensation while passing urine. No bowel complaints. On Examination, the patient was fairly built, pulse was 110 bpm and BP was 110/70mmHg. Abdominal examination revealed tenderness at McBurney's point with no lump and rebound tenderness. Rest of the examination was within normal limits. An emergency abdominal and pelvic ultrasound was performed which revealed a 10 mm dilated non peristaltic blind ending structure in the right iliac fossa with moderate peripheral fat stranding with no faecolith suggestive of Acute appendicitis. Patient was posted for Diagnostic laparoscopy which revealed thickened inflamed appendix with small bowel flimsy adhesions. Careful adhesiolysis was done. Mesoappendix was divided and appendectomy was performed. A tube drain was placed in the pelvis. Patient tolerated the procedure well and was discharged on POD-2 tolerating full diet. On Follow-up examination, the histopathology report revealed an incidentally detected Neuroendocrine tumor of the appendix less than 2 cm involving the tip with free base and mesoappendix invading only up to sub-mucosa and well differentiated Grade 1 morphology.

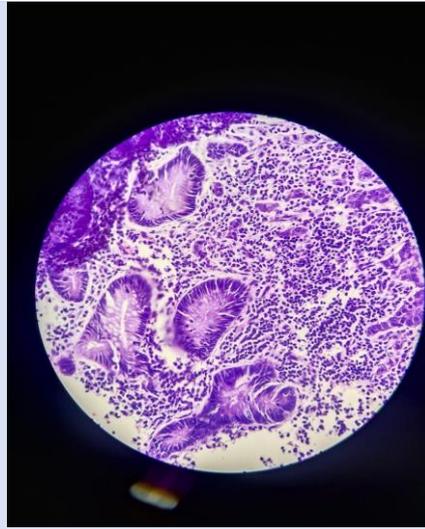


Figure 1: Round or oval nuclei with salt & pepper chromatin & eosinophilic granular cytoplasm in nested pattern.

Discussion

Primary neuroendocrine neoplasia (NEN) has the propensity to develop in any organ of the digestive system. According to the most recent data from the North American Epidemiology Registry (SEER), the most common GI NENs are those arising in the small intestine or rectum, pancreas and stomach or appendix in the order of incidence. Other locations, such as the oesophagus, liver or bile ducts, are exceptional [3]. Neuroendocrine tumor of the appendix is a rare entity [4]. The annual incidence of neuroendocrine tumors of the appendix is estimated to be 0.15 up to 0.6 cases/year per 100,000 people [5]. Their prevalence is estimated to be between 0.3 and 0.9% of appendectomy patients; 95% of appendiceal carcinoid tumors are less than 2 cm in size and 75%

of them are located in the distal third of the appendix [6]. Age of the patient at diagnosis is unique in appendiceal carcinoid tumors, and are often seen in young adults [8]. There are no specific clinical signs or symptoms associated with appendiceal carcinoids. Most NETs of the appendix are asymptomatic and usually discovered incidentally during appendectomy or other benign pelvic surgery. More than 50% of carcinoids mimic acute appendicitis with the classic right lower quadrant abdominal pain, which leads to appendectomy [9]. Carcinoid syndrome is very rare (<1%) [5]. In the vast majority of cases, the diagnosis of appendiceal NET is made incidentally postoperatively in appendectomy specimens that were performed due to either acute appendicitis or recurrent abdominal pain. As most appendiceal NETs are diagnosed postoperatively [10].

Table: Recommended surgical strategies for appendiceal NETs based on specific clinical and histological characteristics.

Indications	Type of operation
Tumor size < 1 cm	Appendectomy
Tumor size 1-2 cm	Appendectomy + Regular F/Up for 5 years
Tumor size > 2 cm	Right hemicolectomy
Location of the tumor at the base of the appendix	Right hemicolectomy
Infiltration of the cecum	Right hemicolectomy
Positive surgical resection margins	Right hemicolectomy
Appendiceal mesentery invasion	Right hemicolectomy
Metastatically infiltrated mesoappendiceal lymph node	Right hemicolectomy
Presence of undifferentiated or low differentiated cells	Right hemicolectomy
Presence of goblet cells	
Goblet cell carcinoma in males	Right hemicolectomy
Goblet cell carcinoma in females (regardless of age)	Right hemicolectomy + Bilateral salpingo-oophorectomy
Peritoneal dissemination from goblet cell carcinoma	Cytoreductive surgery + Adjuvant intraperitoneal chemotherapy

In particular for tumors larger than 2 cm, a CT scan and somatostatin receptor scan (SRS) is recommended at 6 months and 12 months postoperatively and then annually. Colonoscopy is recommended for the early detection of large bowel tumors that are synchronous or metachronously developed [10]. The use of plasma chromogranin-A levels as a tumor marker contributes to the differential diagnosis of calciform cell carcinoma, early detection of recurrence and long-term follow-up of metastatic disease. All patients should be examined 6 and 12 months after surgery and then annually, while follow-up should be lifelong [10]. Hematological investigations, biochemical or urinary investigations are only indicated in rare carcinoid syndrome to monitor disease progression [9].

Conclusion

Post-operative oncological follow-up is not necessary for tumors smaller than 1 cm. On the other hand, it is essential for high-risk tumors and involves an abdominal scan at 3 months post-operatively and then every 6 to 12 months for 10 years. A colonoscopy should also be performed at a distance from the operation to look for a synchronous tumor [4]. Neuroendocrine tumors of the appendix are rare and can be discovered by chance. Its clinical picture is unrecognized, often misleading and non-specific. It is mistaken for acute appendicitis. Although neuroendocrine tumors of the appendix are frequently rare, knowledge of the therapeutic management of these tumors is very important as they have a poor prognosis.

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