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A Carcinoid Tumor of the Cecal Appendix: When an Incidental Finding Dramatically Modifies a Patient´s Prognosis and Treatment

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Abstract

Introduction: Appendectomies are widely used all over the world when there are signs and symptoms suggestive of acute appendicitis and also for prophylactic resection. Study of the surgical specimen is frequent despite the low incidence of unusual findings. Tumors of the cecal appendix constitute 1% of all intestinal neoplasms. Among them, carcinoid tumors are the most frequent. **Objective:** We present a case of a carcinoid tumor of the cecal appendix found incidental to a prophylactic laparoscopic appendectomy during a gynecological procedure. **Case report:** A 58-year-old patient with a history of uterine myomatosis who had had a hysterectomy in 2010 consulted after two years of chronic pelvic pain. An appendectomy was performed because intraperitoneal adhesions from the surgery had affected the cecal appendix. The histopathological analysis identified a typical carcinoid tumor. **Discussion**: Carcinoid tumors are neuroendocrine neoplasms that can be found in various locations but which are most common in the gastrointestinal tract. Involvement of the cecal appendix is not common and is usually detected incidental to prophylactic appendectomies. Identification of this type of neoplasia in a routine study drastically modifies patient management because management and follow-up depending on the tumor's size, extent and location.

Key words

Carcinoid tumor, appendix neoplasia, cecal appendix, appendectomy

INTRODUCTION

Laparoscopic and open resections of the cecal appendix are a widely practiced procedures throughout the world. There are more than 300,000 procedures performed every year in countries like the United States. (1) They are done either as definitive management of acute appendicitis or performed prophylactically at the time of abdominal surgery for another reason. (2)

Histopathological study of the surgically removed tissue is routine, but the infrequent reports of unusual findings in the literature makes its usefulness questionable. (3) A retrospective 14-year study found that of 1,466 appendectomies performed, the percentage of unusual findings was only 3.88%. Of these, neuroendocrine tumors accounted for 0.47% of all cases. (4) Despite the low incidence of abnormal histopathological findings, identifying them leads to changes in treatment and prognosis which makes histopathological study indispensable. (5)

Neoplasms of the cecal appendix account for 1% of neoplasias of the gastrointestinal tract. They can be of various types. Carcinoid tumors are the most common with frequencies ranging from 11% to 50%. Being black, being females and age (peak incidence between the fifth and sixth decade of life) are risk factors. (6, 7)

CLINICAL CASE

The patient was a 58-year-old woman who had a history of hypertension and uterine myomatosis. She who had undergone a hysterectomy in 2010. She came to the primary care unit on a number of occasions because of continuous dull pain in the lower abdomen. Episodes of exacerbation had no apparent triggers or attenuating circumstances and pain was unrelated to any other symptom. Patient claimed to have no other gastrointestinal, gynecological or respiratory symptoms, so her symptoms had been treated for up to two years.

Subsequently, she was referred for gynecological evaluation for chronic pelvic pain. Ultrasound findings showed multiple intraperitoneal adhesions, so she underwent surgery to remove them. Intraoperatively, a general surgeon was consulted because of the large area of intraperitoneal structures, including the appendix, which had been affected. Tissue resected was sent to the laboratory for routine histopathological study. The procedure ended without complications.

When the patient's recovery was complete recovery in terms of surgery and the original complaint, the laboratory's histopathological report was received. It indicated that there was a well differentiated typical carcinoid neuroendocrine tumor of 6 mm diameter in the appendicular tip which had compromised the muscle tissue and blood vessels. Its proximal border was free of tumor. The patient was immediately referred to the oncology department for complementary studies and management.

DISCUSSION

Carcinoid tumors are well differentiated neuroendocrine neoplasms that can be found in the gastrointestinal tract (55%), respiratory tract (30%) and in other locations (15%) such as the kidneys and ovaries. Approximately 45% of those found in the gastrointestinal tract are located in the small intestine while 20% are found in the rectum, 16% in the cecal appendix, 11% in the colon and 7% in the stomach. (8)

Depending on their extent and the principal progenitor cell, carcinoid tumors can produce a large number of neuroendocrine products including insulin, dopamine, gastrin, serotonin, glucagon, vasoactive intestinal peptide, histamine and somatostatin. These act on endogenous receptors and, depending on the amount released, produce specific symptoms. (9)

Facial flushing, usually episodic, is associated with hypotension. It can last between 30 seconds and 30 minutes and may affect the neck and upper thorax. Flushing, secretory-type diarrhea, telangiectasias on or around the nose and cheeks, bronchospasms and even signs of heart failure due to valve dysfunction make up the set of symptoms that constitute the carcinoid syndrome. However, whether or not this syndrome develops depends on the size and of the tumor and degree of metastasis. This is common in neoplasias of the small intestine, although infrequent in carcinoid tumors of the cecal appendix which are usually asymptomatic, as was the case with our patient. (9, 10).

Tumors of the cecal appendix can affect up to 10% of the base of the cecal appendix. Depending on the size and extent of the tumor it may obstruct the lumen, lymphatic flow or venous flow and result in edema and ischemia. When it does, it produces a systemic inflammatory response and the classic symptoms of acute appendicitis. However, the great majority of these neoplasms are detected secondary to prophylactic appendectomies in other surgical procedures, as in the case presented here. (6, 11)

A histopathological study is of vital importance for detection of tumor cells and determination of their histological type, whether resection margins are free of cancer cells, basal or distal compromise of the appendix, tumor size, whether the tumor has invaded various layers of the mucosa, and identification of which layers. All of this is useful for defining definitive therapy which is based more on a consensus of experts than on studies of great statistical power. For those tumors with distal involvement of less than one cm, an appendectomy is usually curative. On the other hand, for tumors larger than two cm, a hemicolectomy is indicated to eradicate metastases to colon and lymph nodes. Treatment of tumors whose sizes are between one and two 2 cm falls into two groups. For tumors with no involvement of the base, an appendectomy is preferred as the only treatment. For tumors with positive borders, invagination of a blood vessel, mixed tumors, and tumors that affect the mesoappendix, treatment is more aggressive and a hemicolectomy is chosen. (6, 10) Depending on the size of the tumor, management is based on the premise and results of studies demonstrating that metastasis is almost nil for tumors smaller than one cm, 7.5% for those measuring between one and two 2 cm, and up to 33% for those larger than two cm. (12)

Prognoses of patients with tumors of the cecal appendix are also affected by size and metastasis. Patients with tumors smaller than two cm that have not metastasized have five year survival rates close to 100%, but patients with tumors whose sizes are between one and two 2 cm which have metastasized to lymph nodes and patients whose tumors are larger than two cm have five year survival rates of about 78%. When the tumor has metastasized to any organ, most commonly the liver, regardless of tumor size, patients' five year survival rate falls to 32%. (13, 14)

Follow-up depends on tumor size. Tumors that are smaller than two 2 cm, for which an appendectomy is considered to be definitive management, do not require follow-up. When a hemicolectomy has been performed for neoplasias

between one and two cm or for tumors larger than two cm, follow-up using abdominal computed tomography (CT) with double contrast, an octreoscan and blood tests to identify markers such as chromogranin are carried out annually to determine whether metastasis to the liver has occurred and to identify any symptoms suggestive of carcinoid syndrome. For patients whose tumor has metastasized distally, follow-up examinations are indicated every 6 months. (15)

Subsequently the patient discussed here was referred to the oncology unit of another institution and is currently undergoing periodic follow-up. To date her tests have been negative for malignancy. We want to emphasize the importance of the histopathological study of the surgical pieces for determining the treatment schemes used for these patients despite the low percentage of unusual findings in these studies.

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