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Case Report

Cancers of the appendix: a case report and review of the literatures

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ABSTRACT

Cancers of the appendix are rare. Most of them are found accidentally on appendectomies performed for appendicitis. Majority of the tumors are carcinoid, adenoma, and lymphoma. Adenocarcinomas of appendix constitute about only 0.08% of all cancers and the treatment remains controversial. Here we are reporting a 57 year old man presented with symptoms of appendicitis, diagnosed with mucinious adenocarcinoma of the appendix. The patient was treated with appendectomy. We performed a review of literature on tumors of the appendix. Benign conditions are treated with surgery alone. For lymphomas chemotherapy are required and carcinoid syndrome can be treated with somatostatin analogues.

Keywords: Appendix cancer, Benign appendix tumors, Malignant appendix tumors

INTRODUCTION

Primary tumours of the appendix are rare malignancies. It may cause appendicitis or rupture of appendicitis which maybe the only first symptom of cancer of appendix. The majority of appendiceal tumors are carcinoids, while the remaining 10% to 20% are mucinous cystadeno-carcinoma, adenocarcinoma, lymphosarcoma, para-ganglioma, and granular-cell tumors. Most common symptoms include acute pain in right lower quadrant and with other symptoms of inflammation like fever, leukocytosis, etc.

During an abdominal surgery if a mass in the appendix is encountered incidentally, an appendectomy is performed. Most masses are benign mucoceles or very small carcinoids which do not require any further management. Chemotherapy or more extensive surgery is indicated if lymphoma or larger carcinoid is identified. Cancer of the appendix is very rare accounting for approximately 0.4% of all gastrointestinal tumors.[1] Most of them are found incidentally on appendiectomies performed for appendicitis, occurring in approximately 1% of appendectomies.[2] Carcinoids being the most common, accounting for approximately 66%, with cystadeno-carcinoma accounting for 20% and adenocarcinoma accounting for 10%.[2]

Other rare forms of cancers include adenocarcinoid, signet ring, non-Hodgkin's lymphoma, ganglioneuroma, and pheochromocytoma. Benign primary processes are mainly mucinous epithelial neoplasms, also called adenomas, cystadenoma, and benign neoplastic mucocele. A retrospective clinicopathologic analysis of appendiceal tumors from 7,970 appendectomies showed 0.9 percent with appendiceal tumors. Acute appendicitis was the most common presentation (49 percent), and 9.5 percent were incidental findings.

Primary malignant tumors of the appendix were found in 0.1 percent of all appendectomies. Secondary malignant disease was identified in the appendix of 11 patients, most commonly (55 percent) from patients with primary colorectal disease. There was a high incidence of synchronous and metachronous colorectal cancer in all appendiceal tumors: carcinoids, 10 percent; benign tumors, 33 percent; secondary malignancies, 55 percent; primary malignancies, 89 percent.[1]

The majority of primary cancers of the appendix occur in 55–65 years of age. Men and women seem to be at equal risk for all appendiceal neoplasms except for malignant carcinoid which may have woman to man ratio in excess of 3:1.[1] All patients with appendiceal neoplasms should be followed because a second malignancy will develop in 15% to 20% of cases.[3]

Carcinoid tumors

Carcinoid tumors were first described over 100 years ago by Lubarsch, who found multiple tumors in the distal ileum of two patients at autopsy. The term *karzinoide* was used by Oberndorfer in 1907 to describe similar tumors that appeared to behave in a more indolent fashion than typical adenocarcinomas.[4] Carcinoid tumors are the second most frequently encountered malignancy of the appendix after adenocarcinoma.[5] Carcinoid tumors most commonly involve the lungs, bronchi, and gastrointestinal tract. Carcinoid tumors are thought to arise from neuroendocrine cells.

They are characterized histologically by positive reactions to silver stains and to markers of neuroendocrine tissue, including neuron-specific enolase.[4] Carcinoid tumors were made up of enterochromaffin cells. These cells produce and contain approximately 90% of the serotonin in our bodies.[6] Majority of carcinoids arise in the appendix,; however, recent data suggests that it may actually be more common in other sites. In a study by Mggard et al 11,427 cases were analyzed, and it was found that 44.7% of tumors were found in the small intestine, 19.6% in the rectum, 16.7% in the appendix, 10.6% in the colon, and 7.2% in the stomach.[7] There is a greater proportion of pulmonary and gastric carcinoids compared to appendiceal carcinoids as suggested from the SEER database.[8]

Carcinoid tumours tend to present with the clinical signs and symptoms of an acute abdomen and frequently coexist with other intestinal neoplasms.[5] It may take around 9 years to become symptomatic.[9] The incidence of carcinoid tumors may vary from one to two in 100,000.[10,11] which may be double in the newer data. Most carcinoid tumors of the appendix are asymptomatic.

When the tumor is located in the tip of the appendix, in approximately 75% of the cases, it generally does not present with symptoms until it becomes metastatic. When the tumor is located at the base of the appendix, it can occlude the lumen and give the patient similar signs and symptoms of appendicitis.[2] In these patients, the diagnosis of carcinoid cancer is typically made by pathology after an appendectomy has been performed.

Rarely patient may present with signs and symptoms related to a carcinoid syndrome like flushing, tachycardia, severe explosive diarrhea, and hypotension. Carcinoid syndrome affects approximately 10% of those with carcinoid tumors. These effects are mostly caused by the serotonin. The carcinoid tumor also produces vasoactive substances such as histamine, prostaglandins, kallikrein, bradykinins, substance P, gastrin, corticotrophin, and neuron-specific enolase. The lungs and the liver are able to clear many of these agents along with the serotonin, therefore, able to avoid carcinoid syndrome. It is not until these organs have carcinoid metastasis that the ability to clear these substances becomes impaired, and symptoms of carcinoid syndrome become apparent.[9]

When the patient presents with signs and symptoms of appendicitis, a CAT scan of the abdomen will usually show a process of acute appendicitis or have associated calcifications.[12,13] In suspected cases of carcinoid syndrome, urinary 5-HIAA and serum chromogranin A level measurements can be useful tool in diagnosis.[9] The extent of surgery is based upon the size of the tumor, but since the majority of carcinoid tumors are found incidentally on simple appendectomies, a second surgery is sometimes needed.

A carcinoid tumor less than one centimeter, or according to some sources between one and two centimeters is appropriately treated with appendectomy alone.[14] The same tumor, measuring greater than two centimeters, or associated with mesenteric/ lymphatic invasion, should prompt right hemicolectomy.[15] Examination of the histological specimen revealing location at the base of the appendix, rather than at the tip also necessitates right hemicolectomy.[14]

Post-operatively, a 3 month follow up should be done with a history and physical examination, CT of the abdomen, and tests for markers (5-HIAA and chromogranin A).[1]

For patients with metastatic disease, somatostatin analogs can be beneficial in relieving the symptoms of carcinoid syndrome. Somatostatin, an 18 amino-acid peptide, binds to somatostatin receptors to block the secretion of hormones such as growth hormones, gastrin, insulin, and glucagon. These receptors are found on over 80% of carcinoid tumors.[16] Octreotide, an eight-amino-acid, long-acting somatostatin analogue, works through G-protein activation on somatostatin receptor subtypes 2, 3, and 5.[17] Octreotide as effective in decreasing symptoms in 88% of patients and decreasing the urinary 5-HIAA in 72% of patients.[8] In patients who do not respond to octreotide, interferon-alpha has been added with some positive

results; however, as stated by Mayer, this therapy comes at a cost of side effects which may include fever, fatigue, anorexia, and weight loss.[8]

Generally, the prognosis for carcinoid tumors of the appendix is very good. If the tumor is confined to the appendix, the disease is said to have 94 percent 5-year survival rate. For patients with regional disease, there is an 85 percent 5- year survival rate, and for distant metastasis, which occurs in approximately 4% of the time in appendiceal carcinoidtumors, there is a 34 percent 5 year survival rate.[11]

Benign appendix tumors

Pathlogically, four different types have been enlisted: Retention cyst, Hyperplastic polyp, Cystadenoma and Cystadenocarcinoma. Like colon cancer, there are both hyperplastic and adenomatous changes at the cellular level.[16]

Hyperplastic polyp

The polyps are sessile or flat and do not have well-defined borders. The involvement is isolated to the mucosa. The obstruction is caused by the hyperplastic polyp.

Cystadenomas

They are the most common neoplasm of the appendix. Much like the Hyperplastic polyp, the growth itself is what causes the formation of a mucocele. Histologically, the lesion is benign. The treatment is surgical excision. Rupture of the appendix can cause pseudomyxoma peritonei.

Retention cysts

They occur when there is a non-malignant obstruction of the outflow tract of the appendix. The most common cause of the obstruction is a fecalith. The epithelial cells continue to produce mucin, and the appendix becomes distended. The vast majority of mucoceles measure less than 2 cm, and those that exceed 2 cm are more likely to be neoplasms.[12,13] Most are clinically asymptomatic and are found incidentally as palpable masses in the lower right quadrant. CT and MRI are more effective in identifying the extent of the mass. Treatment is primarily surgical excision.

Malignant appendix tumors

Adenocarcinoma

Of the appendix is very rare, accounting for 0.5% of all gastrointestinal cancers.[18] Within the adenocarcinoma malignancies there are three subtypes: mucinous (55%), colonic type (34%), and adenocarcinoid (11%) which has a mixed morphology. The mean age of diagnosis is in the fifth decade of life, with an even male to female ratio for all but colonic type, which may have a higher incidence in men.[2] Primary adenocarcinomas of the appendix are very rare malignancy accounting for 0.05-0.2% of all appendectomies and only 6% of all malignant tumors of appendix.[19]

Mucinous adenocarcinoma

Adenocarcinoid, also called Goblet cell carcinoid, has features of both carcinoid tumor and mucinous adenocarcinoma. They account for 5% of cancers of the appendix, with an average age diagnosis of 58 years, and an even distribution between men and women.[2] As is the case with most cancers of the appendix, the most common presentation is that of acute appendicitis.

The mucinous type, also called mucinous cystadeno-carcinoma, causes a mucocele by the neoplasm occluding the narrow lumen which allows the mucin to build up and distend the appendix. Perforation may occur allowing the spill of cancerous cells out into the peritoneum, which creates the condition of pseudomyxoma peritonei. These cells then seed the organs of the peritoneum and continue to produce the mucin. As the mucin accumulates, the abdomen becomes distended which is referred to as "jelly belly".[20] Colonic type adenocarcinoma is less likely to present with a mucocele.

Adenocarcinoma

Adenocarcinoma of the appendix is rarely diagnosed preoperatively. In a study by Nitecki et al, none of the 96 patients with adenocarcinoma of the appendix were diagnosed preoperatively, and it was only considered in the differential diagnosis in 10 patients.

CT imaging is helpful in identifying a mucocele caused by the neoplasm. Calcifications increase the likelihood that this process is malignant. If there is a superinfection associated with the malignancy, there may be air bubbles present on the CT. If the patient presents with a distended abdomen due to pseudomyxoma peritonei, a CT would show widespread heterogeneous locules in the peritoneal cavity.[12,13] But the final diagnosis is most often made postoperatively on microscopic examination.

It has been generally accepted that a right hemicolectomy is the preferred surgical intervention for all subtypes of adenocarcinoma. While some surgeons suggest that a simple appendectomy is sufficient for tumors exhibiting only local disease, many studies have shown that there is a clear survival benefit to the addition of a hemicolectomy.[18,21,22]

In a study by Nitecki, the 5-year survival rate for hemicolectomy was 73% versus 44% in the appendectomy group. These studies have found that the colonic and goblet cell subtypes are invasive, and approximately half the patients present with nodal

metastasis.[18] However, there are some studies that disagree. Gonzalez-Moreno and Sugarbaker found that those patients with mucinous type cancer had no survival benefit from hemicolectomy versus appendectomy.[23]

In a study done by Pahlavan and Kanthanon adenocarcinoitumors, he states that even though Goblet cell carcinoma is an aggressive tumor, a simple appendectomy is appropriate in most cases. However, he further states that a right hemicolectomy should be performed in the following scenarios: (1) cellular undifferentiation, (2) increased mitotic activity, (3) involvement of the base of the appendix, (4) lymph node metastasis, or (5) tumor size greater than 2 cm.[24] These guidelines allow the surgeon some direction in deciding whether or not to reoperate after a patient has had an appendectomy for an apparent appendicitis.

While there are small studies and anecdotal case reports that suggest a response to regimens containing Fluorouracil, the role of chemotherapy has yet to be clearly defined. Most oncologists agree that in the presence of nodal involvement, systemic and intraperitoneal chemotherapy regimens should be used.[18,20,24] Sugarland suggests postoperative intraperitoneal chemotherapy in the setting of pseudomyxoma peritonei, as long as cytoreduction and debulking have been accomplished, reducing recurrence rates.[20]

Prognosis of adenocarcinoma depends on the subtype and extent of disease. Mucinous adenocarcinoma is considered to have a more favorable prognosis because it does exhibit hematogenous or lymphatic spread.[20,21] While it would be natural to assume that those patients with intraperitoneal seeding due to appendiceal perforation, would have a worse prognosis versus those who did not. However, in a study by Nitecki and others, there actually was no difference in 5-year survival rates between the two groups.[18] Perforation often lead to earlier medical intervention and treatment. Goblet cell subtype is considered to have a worse prognosis with one study listing a 55% 5-year survival rate while Pahlavan states a 60-80% 5-year survival rate.[21,24]

Lymphoma

The gastrointestinal tract is the most common site for extranodal lymphoma. The stomach is the most common, followed by the small intestine, pharynx, colon, and esophagus. Lymphoma of the appendix is almost exclusively non-Hodgkin's B-cell lymphoma, more specifically, Burkitt's lymphoma. The incidence of primary appendiceal lymphoma has been estimated at 0.015% of appendectomy specimens.[25] Men are more likely to develop appendiceal lymphoma over women by 1.5:1, with a median age onset of 18 years.[26]

Patients typically present with symptoms similar to acute appendicitis. Patients may also present with a more insidious onset of pain in the right iliac fossa for a few months and an associated palpable mass in the right lower quadrant. When imaging is done, the appendix demonstrates prominent enlargement, while it maintains the vermiform appearance.[12,13] On ultrasound, the diffuse thickening is hypoechoic and often mimics the cystic dilation of the lumen seen in mucoceles.

Chemotherapy is the mainstay of treatment. The classic combination of cyclophosphamide, doxorubicin, vincristine, and prednisone has been used for many decades, and after recent studies, Rituximab has been added.[27] Rituximab, a monoclonal antibody against CD20, was found to increase complete response rates from 64% to 76%.[28] It was also found that the addition of Rituximab increased event free states and overall survival.

Primary signet ring cell carcinoma

Primary Signet Ring Cell Carcinoma of the appendix is a very rare cancer. Based on the data gathered from the SEER database, the incidence is approximately 0.15, with an average age of 59, and there is no male or female preference. Of all the subtypes of appendiceal cancers, signet ring had the lowest percentage of localized disease at 14%, and the highest percentage of distant disease at 60%. The most common presentation is of acute appendicitis, and the recommended surgical intervention is of a right hemicolectomy.[29] The prognosis is considered very poor.[2] Of all the subtypes, signet ring had the lowest overall 5-year survival rate at 18%. Those who presented with distant disease had a 5-year survival of 7%. The role of chemotherapy as a treatment in signet ring cancers of the appendix has not been determined.

Ganglioneuroma

It is an extremely rare cancer that is associated with neurofibromatosis, MEN 2b, congenital defects, carcinomas, and various polyp forming diseases.[17] Only three case reports of ganglioneuromas involving the appendix, all being the Neurofibromatosis type 1.[31-33] In those patients with Neurofibromatosis 1 and MEN 2b, pheochromocytomas are also prevalent and may be found as a primary cancer of the appendix.

CASE REPORT

We are reporting a 57 year old man who presented with acute abdominal pain of 4 days duration; pain was associated with vomiting and low grade fever. There was no history of loss of appetite, weight loss or any symptoms of bowel and bladder involvement, also no past history of similar attack.

Systemic examination was within normal limits but abdominal examination revealed tenderness in right iliac fossa only. Routine investigations like complete haemogram, liver function, kidney function, blood sugar,

X-ray chest and ECG were within normal limits. Ultrasonography of the abdomen showed subacute appendix, retocaecal in origin. Appendectomy was done. This specimen was sent for histopathological examination. On gross examination the appendicectomy specimem measured 6 cm × 0.8 cm in size. Cut section showed dilated lumen filled with small amount of mucus with no lympovascular spread (Figure 1).

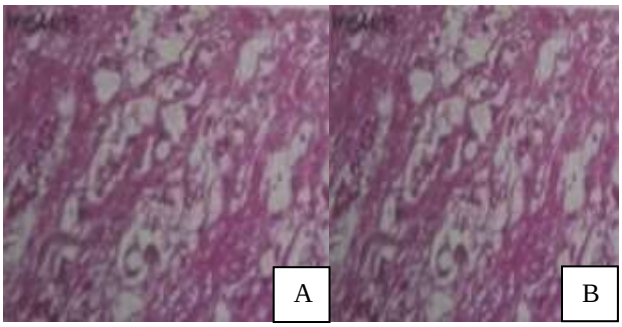

Figure 1: HPE specimen showing malignant tumour composed of signet ring cells. Also seen malignant glands lined by cells with relative bland nuclei in a background of pools of mucin.

Postoperative course was uneventful. Subsequently, he came to radiotherapy clinic for follow up after 6 weeks. The patient underwent staging CT scans of the chest, abdomen, and pelvis with contrast which did not reveal any evidence of residual disease or distant metastases. The patient was offered for adjuvant chemotherapy but patient declined. On further follow up after 4 months, he remained asymptomatic. Thereafter patient got lost in follow up.

DISCUSSION

Appendix cancer is rare, and most commonly an incidentally finding in appendectomies.[9-12] The four main histologic types of appendiceal malignancies are carcinoid tumours, mucinious cystadenomas, adenocarcinomas and adenocarcinoid tumours. For appendiceal carcinoids with tumors larger than 2 cm reoperation and smaller tumors with mesoappendiceal invasion, right colectomy is recommended.[12-15]

Simple appendectomy is a sufficient therapy for benign appendiceal mucoceles, cystadenomas, and some cystadenocarcinomas. A right colectomy is indicated for cystadenocarcinomas with mesenteric or adjacent organ involvement and complicated mucoceles with involvement of the terminal ileum or cecum, cystadenocarcinomas.

Pseudomyxoma peritonei (PMP) is a unique condition characterized by diffuse collections of gelatinous material in the abdomen and pelvis, associated with mucinous implants on the peritoneal surfaces. The natural history is one of indolent but progressive growth, and if left untreated, this is a fatal condition. Standard treatment for PMP is repeated surgical debulking for symptomatic disease. A more aggressive approach using radical surgical cytoreduction of all intraabdominal and pelvic disease and intraperitoneal heated chemotherapy (IPHC) has been adopted by some clinicians, aiming for cure. Five-year survival rates of 70 to 86 percent have been reported for highly selected patients.[18-21]

Adenocarcinomas more often present with a clinical picture of acute appendicitis. Standard treatment is a right colectomy. The role of adjuvant chemotherapy for adenocarcinoma of the appendix is unknown. Oncologists recommend adjuvant 5-Fluorouracil based chemotherapy particularly for patients with node-positive intestinal type adenocarcinoma.[22-28] Selected patients treated with aggressive surgical cytoreduction and IPHC may do well-long term. In uncontrolled series from experienced institutions, long-term survival rates in highly selected patients range from 28 to 72 percent at 3 to 10 years. The benefit of systemic chemotherapy for advanced disease is unknown. Case reports suggest some level of benefit with chemotherapy for mucinous appendiceal adenocarcinomas.[29,30]

CONCLUSION

Present case report and the review of literature are a summary of the tumor of the appendix, and the management based more on opinion of the oncologists and radiation oncologists.

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