Appendiceal Adenocarcinoma with Suppurative Appendicitis: Case Report and Literature Review

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Primary adenocarcinoma of the appendix is rarely encountered and is usually discovered incidentally on pathological examination of surgical specimens. Appendiceal cancer has no distinct pathognomonic symptoms and there are no useful preoperative diagnostic tools available to detect it. We report a case of a 36-year-old man with appendiceal adenocarcinoma with suppurative appendicitis who presented with signs and symptoms of acute appendicitis. Appendectomy was performed through a McBurney's incision. When the pathology report confirmed appendiceal adenocarcinoma, however, further treatment and further surgery were refused. The aim of this report is to review the relevant literature and to increase awareness of appendiceal neoplasms.

Key words: appendix vermiformis, adenocarcinoma, carcinoid, mucinous cystadenocarcinoma, appendicitis

Introduction

Primary noncarcinoid adenocarcinoma of the appendix constitutes less than 0.5% of all neoplasms of gastrointestinal origin¹⁻². Most appendiceal malignancies are diagnosed histopathologically from specimens removed after simple appendicectomy. As there are no specific pathognomonic symptoms for these neoplasms, most are not identified until the disease is advanced. Acute abdominal pain and appendiceal mass are the most common symptoms. Adenocarcinoma of the appendix is rare, however, so its clinical presentation and natural history are not thoroughly understood. Thus, the purpose of this report was to review the relevant literature and to increase awareness of appendiceal neoplasms.

Case Report

An otherwise healthy 36-year-old man visited our emergency room (ER) complaining of abdominal cramping and pain in the right lower quadrant (RLQ) that had persisted for 3 days. The patient did not report any alteration of bowel habits or body weight loss. He described initial peri-umbilical pain that progressed and shifted to the RLQ. On physical examination, neither abdominal distension nor abnormal masses were detected. Rebound tenderness was noted in the right iliac fossa. Rectal examination proved normal. An abdominal plain radiograph showed ileus with fecal impaction. There were no significant findings or abnormal masses noted from computed tomography (CT). Preoperative laboratory testing...
revealed an elevated C-reactive protein (CRP) (6.57 mg/dl), leukocytosis (11,000 /mm$^3$) and normal hemoglobin. Acute appendicitis was highly suspected.

Exploration was performed through a McBurney’s incision. No abnormal sanguinous fluid was detected in the peritoneal cavity. The appendix, which measured 12.5 × 2.0 × 1.6 cm, was acutely inflamed, turgid and thickened. There were also regional edematous bowel loops with dense adhesions. No palpable tumor was identified in the cecum, and the subsequent appendectomy was performed smoothly.

Grossly, the appendix appeared swollen at the tip, with fibrin coating and noted serosal roughness. Examination of the proximal segment revealed stricture with marked wall fibrosis. Microscopically, mild cystic dilatation of the appendiceal tip and consolidation in the remainder of the appendix were also evident. Serial sections of the appendiceal specimen also revealed acute suppuration with transmural neutrophilic infiltrates, mucosal and wall necrosis, as well as thick fibrinopurulent exudates over the serosa. Besides inflammation, diffusely infiltrating signet-ring cell adenocarcinoma was identified from the mucosa to the muscularis propria of the appendix (Fig.1 and 2). The mucin stain was positive. All the tumors were confirmed in the appendiceal wall, showed transiting from the mucosa, with most found in the submucosal region. The serosa and periappendiceal adipose tissue were tumor free.

Postoperative recovery was uneventful, with the patient’s abdominal pain subsiding gradually. When the pathology report confirmed appendiceal adenocarcinoma, however, further treatment and second-look surgery were refused.

Fig. 1 Glandular differentiated tumor cell (black arrow) transiting from the mucosa and infiltrating to the submucosal and muscular wall of appendix. (hematoxylin & eosin stain, x 40)
Discussion

The clinical presentation and natural history of appendiceal adenocarcinoma are not well understood because it is rare. Adenocarcinoma of the appendix has been diagnosed preoperatively\(^3\). However, the tumor is usually found during an appendectomy performed based on signs and symptoms of acute appendicitis.

Epithelial tumors of the appendix have been classified into four distinct types. Carcinoid tumors account for 85% of the epithelial appendiceal tumors followed in order of prevalence, by mucinous adenocarcinoma, colonic adenocarcinoma, and the unusual adenocarcinoid tumors (8%, 4%, and 2% respectively)'(1).

Primary noncarcinoid adenocarcinoma of the appendix is a rare neoplasm with less than 500 total cases reported worldwide\(^8\). In 1993, a retrospective study of 1,740 appendectomies performed over a 10-year period disclosed only 13 primary appendiceal tumors, of which six were adenocarcinomas\(^5\). In Taiwan, just 17 patients with noncarcinoid adenocarcinoma of the appendix were treated at Taipei Veterans General Hospital from 1976 to 1996\(^4\).

In Taiwan, most patients reported with noncarcinoid adenocarcinoma of the appendix were 35-75 years old (median 58), with a peak incidence in the fifth and sixth decades of life\(^4\). A slight male predominance was noted in the literature\(^6\).

The various tumor types differ in their clinical characteristics and biological behavior. Larger tumors have a poorer prognosis, with those under 1cm in diameter unlikely to metastasize\(^2\). Adenocarcinoma originates more commonly in the proximal part of the appendix\(^7\). The site of the appendiceal tumor does not affect the prognosis.
Survival depends mainly on the histological type of the tumor and factors such as the level of invasion, lymph-node and distant metastasis, site of perforation, and the type of operation performed\(^8\). Epithelium tumors, especially adenocarcinomas, have a propensity to spread via the lymph channels.

Adenocarcinoma of the colonic type is highly malignant, with a tendency to metastasize\(^9\)-\(^12\). Mucinous adenocarcinomas of the appendix have a slow growth rate, are rarely spread by the lymphatics, and have a high survival rate after resection\(^13\). Mucinous appendiceal cystadenocarcinomas are usually well-differentiated and tend to produce mucin ascites and gelatinous material associated with an intraperitoneal adenocarcinoma, the so-called "pseudomyxoma peritonei". Metastatic spread has not been shown until late in the disease process\(^14\)-\(^15\). On the contrary, the mucinous and well-differentiated adenocarcinomas of the colon and rectum rarely result in pseudomyxoma peritonei. The characteristic differences that distinguish these adenocarcinomas may be reflected by distinctions at the molecular level. Primary appendiceal adenocarcinomas have high incidences of synchronous and metachronous colorectal cancer\(^1\)-\(^2\). In our case, serial sections of the appendiceal specimen revealed tumor cells with signet-ring differentiation infiltrating through the whole muscularis wall of the appendix. Mucosal involvement with transiting submucosal spreading indicated probable primary lesion. The periappendiceal adipose tissue was tumor free and the regional lymph nodes were free from metastasis.

In conclusion, recognition of carcinoma in the presence of an acutely inflamed appendix is difficult. Intraoperative diagnosis requires a high index of suspicion, careful intra-abdominal examination, frozen sections of all suspicious appendices, and permanent sections of all appendices. Right hemicolectomy is the treatment of choice for carcinoid tumors large than 2 cm in diameter and all adenocarcinomas. More contentious indications include adenocarcinoids with evidence of wall invasion, carcinoids between 1-2 cm in diameter, adenocarinos with spread to the margins, and mucinous appendiceal adenocarcinomas. There is a high incidence of synchronous and metachronous colorectal cancer in all types of appendiceal malignancy.

References

阑尾癌併發阑尾炎病例報告和文獻回顧

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原發性闌尾癌在消化道良性腫瘤中發生率所佔比率不高，比較罕見，不易在早期甚至手術前診斷出
來，大部分案例都在手術標本檢查時才確立診斷；目前尚未有特異性的臨床病理症狀或診斷工具，藉此
在手術前將闌尾癌正確診斷出來。本文報告一例36歲男性主訴右下腹疼痛三天，因局部疼痛合併腹膜炎
症狀，診斷為急性闌尾炎，接受常規的闌尾切除手術，最後病理報告為闌尾癌；回顧相關文獻報告，借
此讓大家在治療急性闌尾炎的同時，必須考慮闌尾癌的發生可能性。

關鍵詞：闌尾，腺癌，類癌，黏液狀腺癌，闌尾炎