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Primary Adenocarcinoma of the Appendix Mimicking Appendicitis: A Case Report

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Abstract: ***Introduction: Adenocarcinoma of the vermiform appendix is a rare neoplasm of the gastrointestinal tract. Presentation mimics acute appendicitis, but right iliac fossa mass and intestinal obstruction have also been reported. These presentations reflect various stages of a locally expanding tumour causing luminal obstruction of appendix. Most of them are found accidentally on appendicectomies performed for appendicitis. Adenocarcinomas of appendix are only 0.08% of all cancers and the treatment remains controversial. Case report: We here present an unusual case of a 70 year old man who presented with right sided abdominal pain for which he underwent a CT scan. This showed a perforated appendix and he was initially treated in the outpatient setting, before having an appendicectomy, the histology of which showed adenocarcinoma of the appendix. He underwent a right hemicolectomy and adjuvant chemotherapy. He has remained well from his carcinoma point of view. Conclusion: Adenocarcinomas of the appendix are a category of rare cancers of the gastrointestinal tract. Although at present they are a well studied pathologic entity, the crucial issue of their preoperative diagnosis remains unsolved.***

Keywords: Adenocarcinoma, Appendix, Perforated appendix, Appendicectomy, Hemicolectomy, Chemotherapy

1. Introduction

Primary tumors of the appendix are unusual and most of them (almost 85%) are carcinoids. Adenocarcinomas of the appendix are a category of rare tumors of the gastrointestinal tract, with a frequency of 0,2% - 0,5% of all intestinal malignancies and 4% - 6% over neoplasmatic lesions of the appendix. The first case of a primary adenocarcinoma of the appendix was reported by Berger on 1882. Mucin-producing cystadenocarcinomas or mucous adenocarcinomas, and non-mucin producing or colonic type adenocarcinomas are included in this category. The main presentation of these tumors is that of an acute appendicitis (30%-50%) or as a palpable mass mainly in the right lower quadrant. Less frequently they may present in female patients as an ovarian tumor. Nevertheless mucous adenocarcinomas are reported as having the greatest tendency among tumors to perforate.

2. Case

The patient is a 70 year old male farmer with a 40 year smoking history. He presented to his GP in January 2011 with right flank pain, which was thought to be renal colic and he underwent an outpatient CT KUB in February 2011. This demonstrated a thickened retrocaecal appendix with an associated inflammatory phlegmon, in keeping with a recent ruptured appendix. On the day of his CT KUB, he was examined by a surgical registrar and was reported to be well, with a soft abdomen and mild right iliac fossa tenderness. His blood tests showed a mild inflammatory response (white cell count 11.3, CRP 119 with otherwise normal U&Es and LFTs). He was discharged home with 1 week of oral Augmentin 625 mg TDS and a CT scan was repeated in a month's time (March 2011). This showed that the phlegmon around the appendix was now smaller, though the retrocaecal appendix was still thick walled, in keeping with partial resolution of the inflammation (see Figure 1).

Figure 1: CTAP – A coronal reconstruction view showing a thickened appendix with surrounding abnormal inflammed tissue

However, clinically, his right iliac fossa pain continued to repeatedly flare up, and he had three further week long courses of oral Augmentin, which only settled his pain temporarily. He was seen in the General Surgical clinic in April 2011, where he found to be still tender and have a palpable mass in his right iliac fossa and thus he was booked for an appendicectomy the same week. During the operation, a retrocaecal appendix was found but extensive adhesions between the caecum and lateral peritoneal wall. The operation was otherwise uneventful and the appendix was removed. Histology subsequently came back as a moderately differentiated mucinous adenocarcinoma of the appendix (T4b N0) with clear margins (see Figure 2).

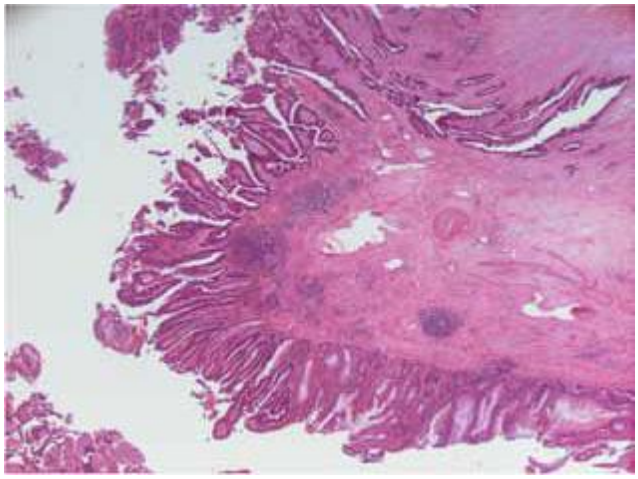

Figure 2: A Haematoxylin and Eosin stained appendix specimen showing a moderately differentiated adenocarcinoma with focal mucin production.

A staging CT of the chest and abdomen showed no metastases and in order to excise the lymph node field, he underwent a right hemicolectomy in June 2011, with an end to end anastomosis, after which he recovered well apart from some post operative ileus which settled spontaneously. Following this, it was planned that he would have 6 cycles of adjuvant chemotherapy with Oxaliplatin and Capecitabine. The patient took all the 6 cycles of chemotherapy and now presents to the hospital with full recovery.

3. Discussion

The most common primary appendiceal neoplasms are carcinoid tumours, comprising around 32%-85% of all appendiceal tumours[1,2]. Primary adenocarcinomas of the appendix, on the other hand, make up only around 4%-6% of all primary appendiceal neoplasms [3] and account for just 0.4%-1% of all gastrointestinal malignancies [4].Only about 250 cases of primary adenocarcinoma of appendix have been described since Berger first recognized the neoplasm in 1882. The mean age of presentation is in the fifth or sixth decade [5], and the incidence of these neoplasms is 0.12 cases per million people per year [6], making them rare. Primary adenocarcinomas of the appendix can be of three subtypes: mucinous (55%), colonic type (34%) and adenocarcinoid (11%) [7], and most neoplasms are only identified after histological examination [8], being found in approximately 0.9%-1.4% of all appendicectomy specimens.

Primary adenocarcinomas of the appendix commonly present as acute appendicitis or with a palpable right iliac fossa mass [9], but rarer presentations include urinary frequency mimicking bladder cancer [10], hydronephrosis [11], mimicking Crohn's disease [12], caecal intussusceptions [13] and anaemia [14]. Primary adenocarcinomas of the appendix are the most frequently perforating carcinoma of the gastrointestinal tract [15] and like carcinoma of the colon, spread via local invasion, lymphatic vessels, and the bloodstream to the peritoneal cavity most commonly, followed by lymph nodes, liver, ovaries, the abdominal wall and the lungs [16].

Table 1: Characteristics of patients presenting with appendiceal adenocarcinoma

Characteristics	*Mucinous adenocarcinoma*	*Colonic type adnocarcinoma*	*Signet ring cell carcinoma*
Age at diagnosis	60 (Range 17–99)	62 (Range 19–98)	58 (Range 25–90)
Gender	Male 49% Female 51%	Male 60% Female 40%	Male 46% Female 54%
Race	White 89% Black 6% Other 5%	White 80% Black 13% Other 7%	White 93% Black 1% Other 4%

The preferred surgical treatment is still controversial. While simple appendicectomy seems to be sufficient for early, non-invasive carcinomas of the appendix [4], most tumours usually present as advanced invasive carcinomas and secondary right hemicolectomy is usually recommended as the operative treatment of choice [17]. The role of adjuvant chemotherapy in adenocarcinomas of the appendix is not clear. Despite this, many oncologists recommend adjuvant 5-fluorouracil based chemotherapy particularly for patients with no depositive intestinal type adenocarcinoma. Cardiotoxicity and colitis are two rare but serious complications of 5-fluorouracil therapy [18,19].

4. Conclusion

Mucin producing adenocarcinomas of the appendix are a category of rare cancers of the gastrointestinal tract. Although at present they are a well studied pathologic entity, the crucial issue of their preoperative diagnosis remains unsolved. Appendicular lesions, both inflammatory and neoplastic, are notorious for atypical presentation. It is thus not surprising that the rate of this misdiagnosis is quite high particularly if solely based on clinical grounds.

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