Primary Adenocarcinoma of Appendix Presenting as Omental Metastasis

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Abstract
The adenocarcinoma of appendix is a rare entity. The patients present usually with symptoms of appendicitis, a palpable abdominal mass or rarely as a complication of pseudomyxoma peritonei. We report a case who presented with lump abdomen and ascitis and revealed a tiny infiltrative growth at the base of appendix at laparotomy.

Key Words
Adenocarcinoma, Mucin, Appendix, Pseudomyxoma peritonei

Introduction
The tumours of appendix are infrequent and usually found after appendicectomy (1). The malignant tumours consist of carcinoid, mucocele and adenocarcinoma. The primary adenocarcinoma of the appendix is extraordinarily a rare tumour. These tumours may arise in pre-existing cystadenomas but rarely in conventional adenoma (2). The 5 years survival is 18.7% for all patients (3).

Case Report
A 40 year old male patient presented with history of gradually increasing pain and swelling in the region of umbilical, hypogastric and right iliac fossa, over a period of two months. Slight weight loss and anorexia was also present. No history of bowel disturbance, melena or jaundice was reported. Examination revealed 10 × 8 cms soft midline swelling in abdomen. Mild fluid thrill was noted. Barium meal follow through was unremarkable. Ultrasound revealed a matted mass with ascitis and suggested metastatic deposits.

At laparotomy, whole of gastrointestinal tract (GIT) was seen normal except a 2 × 1 cms firm growth at the base of appendix involving adjacent part of cecum and infiltrating the wall. The omentum was studded with pearly white nodules, 1.5-2 cms. in diameter. The peritoneal cavity contained about 300 ml. of mucinous fluid. Right hemicolecotomy and meticulous removal of mucus and peritoneal implants was carried out.

Grossly, the specimen was 25 cms long consisting of parts of right colon, cecum, appendix and a part of terminal ileaum. A 2 × 1 cms growth was seen at the base of appendix involving adjacent part of cecum and infiltrating the wall through and through. No serosal deposits or lymph nodes were seen. The appendix was 3 cms long with its lumen filled with mucin but no luminal dilatation was seen. The omentum was flat, 1.5 × 1 ft. in dimensions and was studded with regularly spaced pearly white nodules, 1.5-2 cm diameter. Cut section of the nodules showed partly solid and partly mucoid surface.
The microscopic examination of the main tumour from appendix showed histological features of well differentiated adenocarcinoma with evidence of mucin production. The sections from omental tissue showed deposits of similar well differentiated adenocarcinoma with pools of mucin in which many malignant glands and tumour cells were seen floating. The micro-dissection of the specimen did not reveal any lymph node metastasis.

Fig. 1. Microphotograph showing glands of well differentiated adenocarcinoma infiltrating muscle layer of appendix. (x400X).

Fig. 2. Microphotograph showing pools of mucin with malignant cells in glands from nodular deposits in omentum. (x400X).

Discussions

The tumours of the appendix are infrequent and when found, they are almost always during a routine appendicectomy. In a study of 71,000 specimens removed at appendicectomy, Collin observed an incidence of 4.6% for benign and 1.3% for malignant tumour (1). The malignant tumours of appendix consist of carcinoid, mucocele and adenocarcinoma. The remaining including sarcomas are very rare tumours (3).

The primary adenocarcinoma of the appendix is extraordinarily rare tumour with less than 300 cases described so far in the collected literature (4). The incidence varies from 0.004% reported by Norment (2) to 0.08% reported by Collin (1).

The tumour is most common in persons aged 51-55 years and present either with episode of acute appendicitis or as mass in right iliac fossa. Very rarely, the first symptom is that of metastasis and complications of pseudomyxoma peritonei (3).

These tumours may arise in pre-existing cyst-adenomas but rarely in conventional adenoma and form a papillary or tubular adenocarcinomas without abundant mucin production. The adenocarcinomas arise most often near to the base of the organ. The exact site of origin is the junction between cecum and appendix (5). The histological features are those of large intestine adenocarcinoma. The mucinous tumours tend to infiltrate through the wall of appendix and produce multiple deposits of tumour on peritoneum—the malignant myxoma peritonei. Rupture of the pseudomyxoma cysts also leads to spilling of mucus and epithelium into the coelomic cavity in nearly 2-5% of the cases (3). The tumour cells proliferate in nodular fashion on peritoneal surface and continue to secrete mucin. Such cases have poor prognosis (3).

The adenocarcinomas behave aggressively with frequent metastasis. About 30% of the well differentiated adenocarcinomas have metastasized as compared to 70% poorly differentiated carcinomas at the time of presentation (3).
Right hemicolectomy with resection of the draining lymph nodes is the treatment of choice. Despite the availability of appropriate treatment, survival remains low, 5 years survival with Dukes stage A lesion is 94%, for stage B is 83% and for stage C is 44% only (3).

References


