Management of an Appendiceal Mass
- Approach to acute presentation of appendiceal neoplasms

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Case Scenario

- M/42; unremarkable past health
- Periumbilical pain shifted to right lower quadrant for two days
- Associated with nausea and subjective account of fever
- Physical examination:
  - Temperature 38.9°C; Stable hemodynamics
  - Localized tenderness and “fullness” over right lower quadrant
Suspected Appendiceal Neoplasm

- A CT scan was performed
- Complex cystic appendix with thickened wall
- Suspicious of underlying neoplasm
- Tachycardia, persistent tenderness and pyrexia despite antibiotic therapy
Introduction

1. Overview of commonest appendiceal neoplasms and mucocele

2. Management of acutely presenting appendiceal mass suspicious of underlying neoplasm
Differential diagnoses

- Neoplasms of vermiform appendix
- Secondaries from urogenital tract (ovaries), large bowel, lung and breast
- Others (rare): ganglioneuroma, pheochromocytoma, mesenchymal tumors, Kaposi sarcoma
- Appendiceal mucocele

Table 1. Distribution of Appendiceal Tumors

<table>
<thead>
<tr>
<th>Type</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carcinoid</td>
<td>42</td>
</tr>
<tr>
<td>Benign tumors</td>
<td>12</td>
</tr>
<tr>
<td>Mucinous cystadenoma</td>
<td>7</td>
</tr>
<tr>
<td>Villous adenoma</td>
<td>5</td>
</tr>
<tr>
<td>Malignant tumors</td>
<td>20</td>
</tr>
<tr>
<td>Primary</td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma</td>
<td>8</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>1</td>
</tr>
<tr>
<td>Secondary</td>
<td>11</td>
</tr>
<tr>
<td>Total</td>
<td>74</td>
</tr>
</tbody>
</table>
Appendiceal Neoplasms & Mucoceles
Appendiceal Carcinoid

- Neuroendocrine origin, arise from primitive stem cell
- Classified according to location of primitive gut
- Deep in mucosa for intestinal carcinoids
- Appendix is the commonest site for carcinoid tumor
- Carcinoid is the commonest primary neoplasm of the appendix
Appendiceal Carcinoid

- 0.3-0.9% appendicectomies; usually asymptomatic, incidental finding in appendicitis
- Most affect distal 1/3; invade wall
- Symptoms occur with metastasis (rare)
- Liver metastases are rare; Lymphatic spread as 1* route
- Worse outcome with large size (>1.5-2cm), goblet cell carcinoid (adenocarcinoid)
Intestinal and Mucinous Neoplasms

- Benign: adenomas, cystoadenomas
- Malignant: adeno/ cystoadenocarcinomas
- More goblet cells in appendix than in colon
  -> majority of neoplasms are mucinous in nature
Cystoadenocarcinoma (mucinous)

- Appendiceal carcinomas: 90% mucinous (vs intestinal type)
- *Mucinous* subtype:
  - Tip or along lumen
  - Mucocele formation
  - Dissemination to peritoneal space, pseudomyxoma peritonei
Adenocarcinomas (intestinal)

- Appendiceal carcinomas: 10% intestinal type
- **Intestinal** subtype:
  - Appendiceal orifice
  - Lymph node spread
Appendiceal Mucoceles
A rare but not entirely separate entity

- Morphological description

- Cystic dilatation of appendix caused by accumulation of mucus secretion

- 0.2-0.4% appendicetomy specimens

- Course and prognosis depends on histological subtype


- Often asymptomatic (51%); abdominal pain (27%), mass (16%), weight loss (10%), appendicitis (8%)

- In the presence of pseudomyxoma peritonei or mucinous dissemination, 80-90% are malignant

![Histological Subtypes of Appendiceal Mucoceles]

- Mucinous cystadenomas (52%)
- Mucosal hyperplasia (20%)
- Simple mucoceles/retention cysts (18%)
- Mucinous cystadenocarcinomas (10%)
Management of Suspected Appendiceal Neoplasms
1. Emergency Surgery

- Detailed laparoscopy
  - Macroscopic features of index lesion:
    - Location on appendix, caecal involvement
    - Mesoappendiceal involvement
    - Size of lesion (>2cm)
    - Any mucinous dissemination or spontaneous perforation
    - Any other primary lesion (appendiceal secondary?)
1. Emergency Surgery

- Pathology and histological characteristics of appendiceal tumor determine extent of definitive surgical resection and need of prophylactic lymph node removal

- Achieve macroscopic resection margin, including mesoappendix
  - Appendicectomy, ileocaecotomy, right hemicolecotomy?

- Avoid trauma or rupture during removal

- Use of endobag; retrieval from midline (facilitate port site removal if necessary)

- Thorough peritoneal washout if mucinous dissemination

- Wait for histology result and consider second operation
2. Further Investigations?

- **Non-carcinoid neoplasms:**
  - Tumor markers: CA-125, CA-19-9, CEA (in mucinous peritoneal carcinomatosis)
  - Computed tomography: look for distant metastasis, serve as baseline for disease monitoring

- **Carcinoid neoplasms:**
  - Serotonin metabolite 5-HIAA 24-hour urine sample; chromogranin A (lower specificity)
  - Nuclear imaging: OctreoScan (somatostatin analogue) (80% carcinoid with somatostatin receptor); metaiodobenzylguanidine (MIBG) scan
  - PET scan: based on metabolism of tryptophan

Surveillance colonoscopy: **ALL** appendiceal neoplasms are associated with synchronous or metachronous colonic neoplasms
3. When to Consider Completion Right Hemicolecctomy?

- Prophylactic resection for occult lymph node metastasis
- Based on histolopathological findings of initial resection

- **Carcinoid**: right hemicolecctomy is recommended for lesions >2cm
  - Increased probability of nodal involvement 30% (<0.1% for lesions <1cm) (Sutton et al); controversy for lesions between 1-2cm
  - Involvement of mesoappendix, atypical microscopic foci, mitotic count of 2 or more per HPF, goblet cell type
3. When to Consider Completion Right Hemicolecctomy?

- **Adenocarcinoma**: tradition approach is to offer right hemicolecctomy

  - Much higher incidence of lymph node metastasis in adenocarcinoma (66.7%) than mucinous carcinoma (4.2%) (Gonazlex Moreno et al)

  - >2cm, poor differentiation, lymphovascular permeation, submucosal invasion

- **Mucinous adenocarcinoma**: more selective approach to right hemicolecctomy

  - Median survival similar for those with or without lymph node involvement (28 vs 26 months) (Kiran at al)
3. When to Consider Completion Right Hemicolecctomy?

• Presence of mucinous dissemination:
  
  • Low grade mucinous neoplasms: no recurrence in 6-year follow-up
  
  • vs those with extra-appendiceal spread: 45% 5-year survival
  
  • *Radical resection does not alter prognosis of mucinous neoplasm with peritoneal dissemination*
  
  • Not advisable unless intraperitoneal chemotherapy and cytoreductive surgery
4. Adjunctive Therapy?

- **Carcinoid neoplasms:**
  - Metastatic carcinoid: 5-fluorouracil (5-FU) and leucovorin
  - Hepatic metastases: resection/ablation, interferon, hepatic artery embolization (>50% involvement), long-acting somatostatin analogue (carcinoid syndrome)

- **Non-carcinoid neoplasms:** NO controlled study
  - Same regime as colonic adenocarcinoma
  - Poor tumor response in case of peritoneal dissemination; intraperitoneal heated chemotherapy
Management Algorithm for Suspected Appendiceal Neoplasm

- Acute appendiceal mass (suspicious CT findings)
  - Failed antibiotic therapy

- Emergency resection (macroscopic margin + mesoappendix)
  - Histology proven neoplasm

- Further investigations (tumor markers, imaging, colonoscopy)

- Determine need for completion right hemicolecotomy

- Specialist centre referral/Oncologist referral/Surveillance
Conclusion

- Appendiceal neoplasms are rare but often present as acute appendicitis
- Most common lesions are carcinoid neoplasms & adenoma / adenocarcinoma of intestinal and mucinous types
- Initial treatment without prior histological diagnosis proves a challenge
- Achieve macroscopic margin with careful specimen handling
- Option of a planned second operation is available
- Follow up management including surveillance colonoscopy is necessary
References


- Whitefield et al. Surgical management of primary appendiceal malignancy. Colorectal Dis 2012; 14, 1507-1511