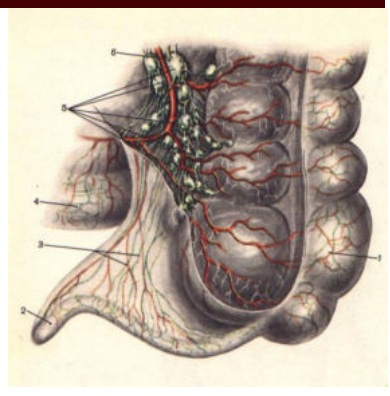
Appendicular Tumors



Neoplasms of the appendix are not often suspected before surgery and are found either intraoperatively or on pathologic examination

Historical background

1842 Rokitansky recognizes the first mucocele of the appendix.

1884 Werth describes gelatinous material in the peritoneal cavity, and attributes it to an ovarian cyst.

1901 Fraenkel reports finding pseudomyxoma peritonei (PP) in a male patients due to a ruptured cyst of the appendix.

1940 Woodruff and McDonald at Mayo Clinic classify 146 mucinous cystic tumors of the appendix.

- "Benign mucocele"
- Cystadenocarcinoma, grade 1 (malignant mucocele) – Papillary arrangement of the mucosa with hyperchromatic elongate nuclei. Only these can cause PP.

In the 1950s, 60s, and 70s, doubts emerged about the malignant nature of these tumors when confined to the appendix. Since they resembled colonic adenomas, several authors used the term "mucinous cystadenomas" or "villous adenomas" to describe these tumors.

Appendiceal tumours can be broadly classified as:

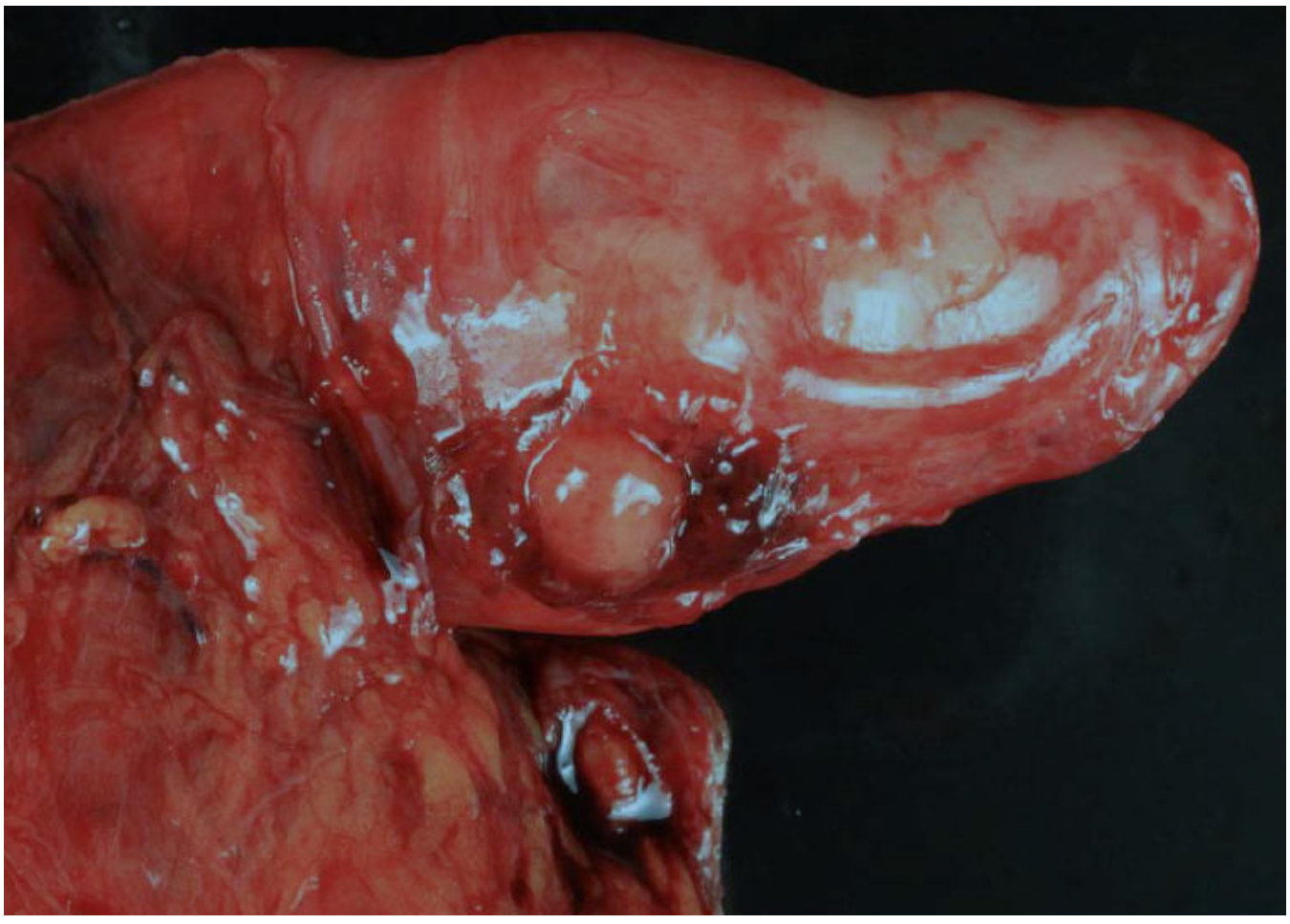
- Epithelial

- Nonepithelial tumors

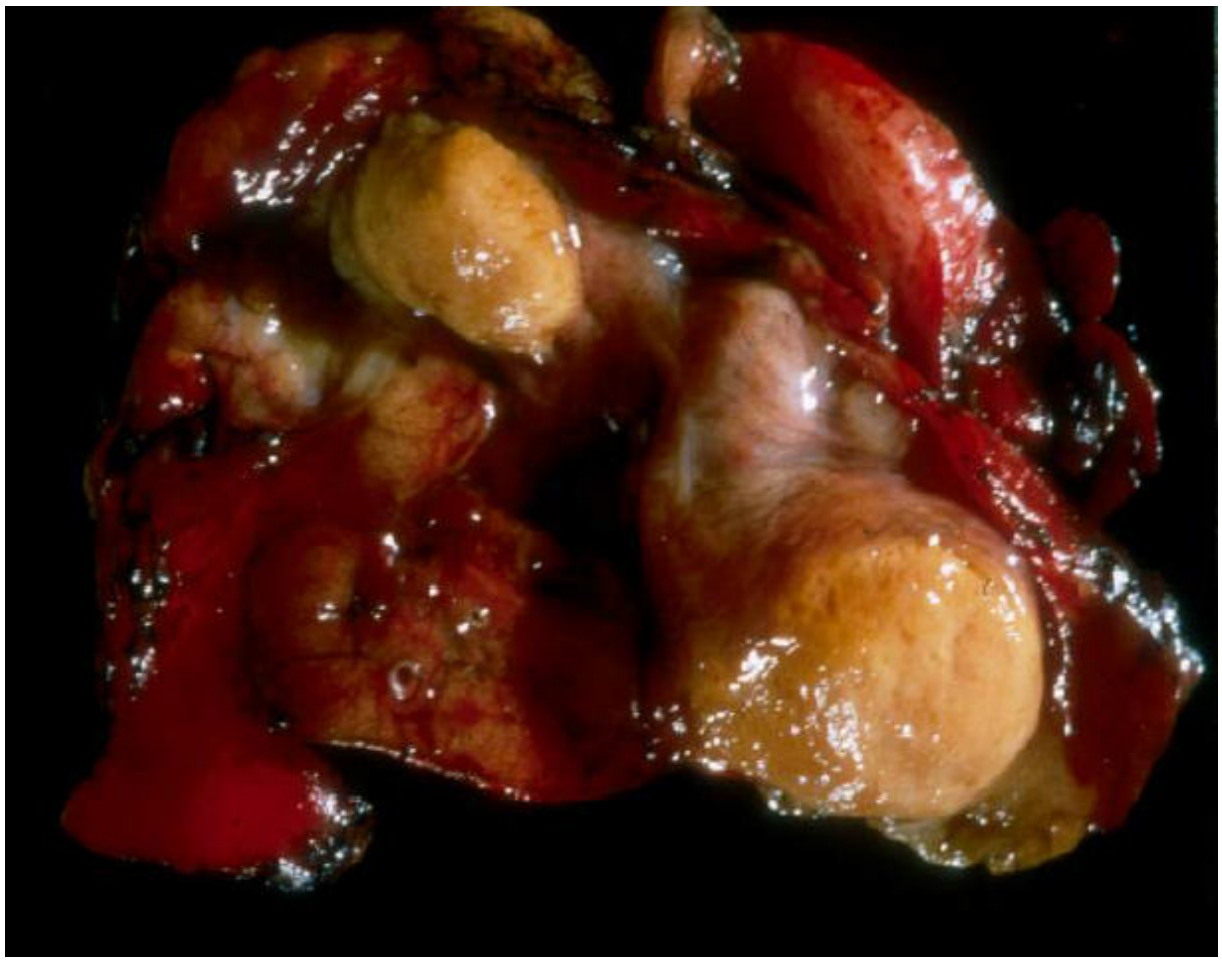
Epithelial tumors

- Adenoma: mild-to-moderate atypia, mitosis, no stromal invasion, perforation with mucin
- Mucinous tumor of uncertain potential: adenoma with positive margin, mucin present within the wall
- Mucinous tumor–low malignant potential: adenoma with neoplastic cells in peritoneum
- Adenocarcinoma: invasive mucinous tumor

Low grade appendiceal Mucinous Neoplasm



Carcinoid tumor



Nonepithelial tumors

Endocrine tumors

- Classic appendiceal endocrine tumors
- Goblet cell carcinomas

Lymphoma

Sarcoma

Carcinoids

MC located at the tip of the appendix

Two types:

- insular type
- tubular

<u>Management</u>

- Tumors ≤1 cm: appendectomy.
- 1 cm to 2 cm (without involvement of the base of appendix): appendectomy

Right hemicolectomy depends on grade, mitotic activity, invasion of mesoappendix, or lymphovascular invasion.

- ≥ 2 cm are at risk for lymph node or distant metastasis and a right hemicolectomy is indicated.

Epithelial tumors

Presentation:

- Incidental finding in the appendectomy specimen
- Appendicitis
- Pelvic mass
- Peritoneal carcinomatosis with or without ascites.

Diagnosis

Biochemistry

Chromogranin A can be used as tumour marker in appendiceal endocrine tumours and is useful to differentiate the tumour from goblet cell carcinoids. It is indicated in metastatic disease as a biochemical parameter for follow-up

CT scan

Histopathology

Staging

Grade of epithelium beyond appendiceal mucosa

Low-grade mucinous carcinoma

High-grade mucinous carcinoma

Tumor stage

T1: Tumor involves submucosa

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T2: Tumor invades muscularis propria

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T3: Tumor invades subserosa or mesoappendix

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T4a: Tumor penetrates serosa, including tumor in the right lower quadrant

Distant metastases

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M1a: Intraperitoneal metastases beyond right lower quadrant

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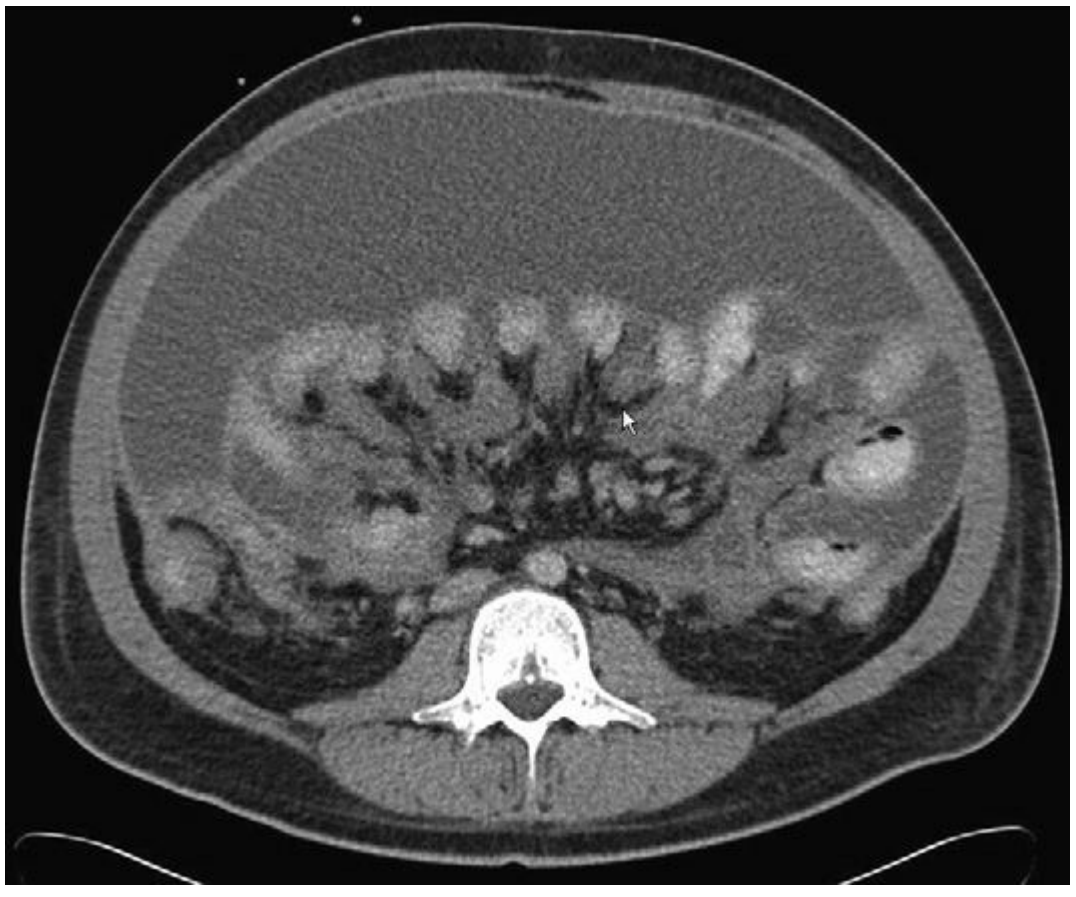
M1b: Extra-peritoneal metastases

Pre operative CT scan



Treatment

Surgery

- Appendiceal tumours can be cured by appendectomy if the tumour is located at the tip of the appendix, the tumour diameter is <2 cm and no deep meso appendiceal invasion is observed

- Right hemicolectomy, is indicated if:
  - tumour diameter >2 cm
  - deep mesoappendiceal invasion
  - positive surgical margins

Medical therapy

- No medical therapy is indicated in patients with resected appendiceal endocrine tumours.
- Chemotherapeutic options are not available on an evidence-based level, nor are data to recommend peptide radioreceptor therapy (PRRT).
- PRRT may be an option in a somatostatin receptor-positive, metastasized, inoperable appendiceal endocrine tumour.

Systemic chemotherapy in appendiceal tumor

- Preoperative (neoadjuvant)
- Postoperative (adjuvant)
- Postoperative after suboptimal cytoreduction with residual bulky disease
- Palliative in unresectable or progressive and metastatic disease

Minimal Consensus Statement on Diagnostic Procedures for Follow-Up

- For well-differentiated tumours, diagnosed incidentally, with a maximum diameter < 1 cm and R0 resection, no follow-up is required.

- For well-differentiated tumours of 1 to < 2 cm and R0 resection there are no sufficient data for a clear-cut decision.

- In cases with deep mesoappendiceal infiltration or angioinvasion, CT of the abdomen and somatostatin receptor scintigraphy may be performed.

- Factors believed to argue for follow-up investigations are a high proliferation marker, vascular involvement, deep mesoappendiceal infiltration, and possibly location at the base of the appendix