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## Vermiform Appendix Tumours

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### I. GROSS DESCRIPTION

#### **Specimen**

- appendectomy/right hemicolectomy. Usually because of acute appendicitis, an inflammatory appendix mass or as part of a colectomy for other reasons, e.g. colonic cancer. Also in the context of ovarian cystic tumours.
- length and diameter (cm).
- mucocoele/perforation/diverticulum/appendicitis/appendicular mass.

#### **Tumour**

#### **Site**

- tip/base/diverticulum/body.

#### **Size**

- length × width × depth (cm) or maximum dimension (cm).

#### **Appearance**

- polypoid/sessile/plaque/ulcerated/infiltrative/mucoid/yellow.

#### **Edge**

- circumscribed/irregular.

### 2. HISTOLOGICAL TYPE

#### ***Carcinoid (endocrine cell) tumours***

- 0.5–1.5% of appendectomies.
- 85% of appendiceal tumours.
- usually a coincidental finding of yellow pale tumour at the tip, although it may contribute to appendicitis when at the appendix base (10%).
- variably chromogranin, synaptophysin, CD56 positive depending on EC (usual) or L cell origin.

*Usual type:* 70% of cases and at the appendiceal tip. Solid nests/cords/ribbons/acini of uniform cells often with invasion of muscularis,

± serosa and lymphatics. Benign with appendicectomy the treatment of choice.

Rarely cases spread to peritoneum, regional nodes and liver (35% 5-year survival) and these are usually >2 cm diameter with size the main factor predictive of behaviour. Radical surgery should be considered in these circumstances. Extensive invasion of mesoappendix and the appendiceal base are also adverse indicators.

*Goblet cell carcinoid (mucinous/adenocarcinoid/crypt cell carcinoma) type:* clusters, strands or glandular collections of mucus-secreting epithelial cells often with a signet ring or goblet cell morphology. Usually only a minor population of endocrine cells is present demonstrated by immunohistochemistry. Potential for extra-appendiceal spread (20% of cases) and occasionally involvement of regional nodes and liver. Propensity for transcoelomic spread to involve ovaries and direct spread through the appendix base and into the caecum.

Right hemicolectomy should be considered, particularly if there is extensive spread with an infiltrative growth pattern, involvement of the appendix base or tumour pleomorphism and mitoses. The term mixed carcinoid/adenocarcinoma is sometimes used if there is an infiltrating component of colorectal type tumour tissue and again radical surgery should be considered.

Distinguish from: (1) secondary colorectal carcinoma involving the appendix either directly (e.g. from caecal pouch) or via the peritoneum (signet ring cell carcinoma of rectosigmoid area), and (2) primary colonic-type mucinous adenocarcinoma of appendix, which is aggressive in behaviour and requires radical surgery. A pre-existing mucosal adenoma in a carcinoma and the component of endocrine cells in a carcinoid lesion may help in this respect.

### **Adenoma**

- <1% of appendicectomies and the majority are benign.
- localized (polypoid—rare) or diffuse.
- tubular/tubulovillous/villous\* with variable grades of dysplasia.
- \*see Mucocoele (Appendiceal Mucinous Neoplasms).
- the adenomatous mucosa can be flattened and simplified, or serrated.
- in up to 20–40% of cases there are adenomas or adenocarcinomas elsewhere in the colorectum.

### **Adenocarcinoma**

- 0.1% of appendicectomies.
- requires destructive invasion through the muscularis mucosae by malignant glands (sometimes but often not with a desmoplastic reaction), or the presence of epithelium in extra-appendiceal mucus (cytokeratins and CEA can be useful in demonstrating this). The latter can be difficult to distinguish from a LAMN with mucin dissection and peritoneal spillage (see below).
- identified as primary by a mucosal adenomatous lesion.