Tumors of Appendix

Dept of Surgery

Introduction

- Appendiceal malignancies are extremely rare.
- Diagnosed in 0.9-1.4% of appendectomy specimens.
- Carcinoid is the most common appendiceal malignancy
- It represents more than 50% of the primary lesions of the appendix.

WHO classification of Appendiceal tumors

Epithelial tumours		Non-epithelial tumours	
Adenoma	8140/0*	Neuroma	9570/0
Tubular	8211/0	Lipoma	8850/0
Villous	8261/0	Leiomyoma	8890/0
Tubulovillous	8263/0	Gastrointestinal stromal tumour	8936/1
Serrated	8213/0	Leiomyosarcoma	8890/3
		Kaposi sarcoma	9140/3
Carcinoma		Others	
Adenocarcinoma	8140/3		
Mucinous adenocarcinoma	8480/3	Malignant lymphoma	
Signet-ring cell carcinoma	8490/3		
Small cell carcinoma	8041/3	**Secondary tumours**	
Undifferentiated carcinoma	8020/3		
		Hyperplastic (metaplastic) polyp	
Carcinoid (well differentiated endocrine neoplasm)	8240/3		
EC-cell, serotonin-producing neoplasm	8241/3		
L-cell, glucagon-like peptide and PP/PYY producing tumour			
Others			
Tubular carcinoid	8245/1		
Goblet cell carcinoid (mucinous carcinoid)	8243/3		
Mixed carcinoid-adenocarcinoma	8244/3		
Others			

Carcinoid

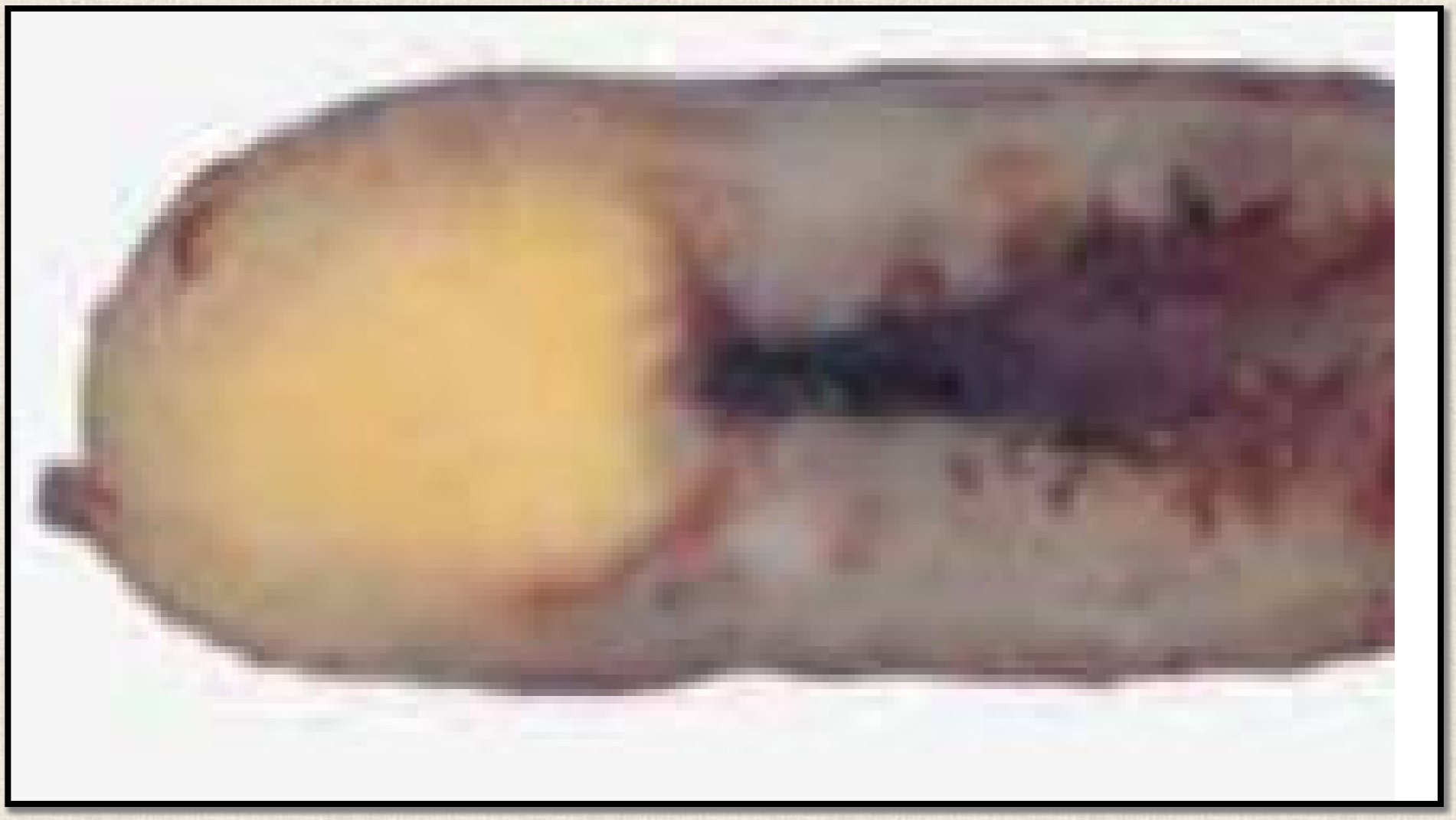
- Most common site: Jejunum>Appendix > rectum
- Mean age at presentation is 32-43 years (range-6 to 80 years)
- More frequently in females than in males
- Carcinoid syndrome is rarely associated with appendiceal carcinoid
- Symptoms attributable directly to the carcinoid are rare
- Tumor can occasionally obstruct the appendiceal lumen and result in acute appendicitis

Macroscopy

- Firm, greyish-white (yellow after fixation), fairly well circumscribed, but not encapsulated
- Measure usually less than 1 cm in diameter
- Tumours > 2 cm are rare
- Most are located at the tip of the appendix
- Goblet-cell carcinoids and mixed endocrine-exocrine carcinomas of the appendix may be found in any portion of the appendix.

Carcinoid

- Tumours with endocrine differentiation arising in the appendix
- Majority of carcinoids are located in the tip of the appendix.
- Malignant potential is related to size
- Treatment :
  - carcinoid tumor is localized to the appendix: simple appendectomy
  - tumors <1 cm with extension into the mesoappendix and tumors >1.5 cm: right hemicolectomy



Carcinoid tumour of appendix with typical yellow colouration

Adenocarcinoma

- Primary adenocarcinoma: rare neoplasm
- Three major histologic subtypes:
  - Mucinous adenocarcinoma
  - Colonic adenocarcinoma
  - Adenocarcinoid

TNM Staging

TNM classification[1, 2]

T – Primary Tumour

TX	Primary tumour cannot be assessed
T0	No evidence of primary tumour
Tis	Carcinoma in situ: intraepithelial or invasion of lamina propria[3]
T1	Tumour invades submucosa
T2	Tumour invades muscularis propria
T3	Tumour invades through muscularis propria into subserosa or into non-peritonealized periappendiceal tissue
T4	Tumour directly invades other organs or structures and/or perforates visceral peritoneum

N – Regional Lymph Nodes

NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Metastasis in 1 to 3 regional lymph nodes
N2	Metastasis in 4 or more regional lymph nodes

M – Distant Metastasis

MX	Distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

Stage Grouping

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
	T2	N0	M0
Stage II	T3	N0	M0
	T4	N0	M0
Stage III	Any T	N1	M0
	Any T	N2	M0
Stage IV	Any T	Any N	M1

- Most common mode of presentation is of acute appendicitis.
- Ascites
- Palpable mass
- Incidental intraoperative finding

Treatment:

- Right hemicolectomy

Appendiceal adenocarcinomas have a propensity for early perforation

Overall 5-year survival is 55%

Patients have significant risk for both synchronous and metachronous neoplasms

Mucocele

Progressive enlargement of the appendix from the intraluminal accumulation of a mucoid substance.

Mucocele are of four histologic types

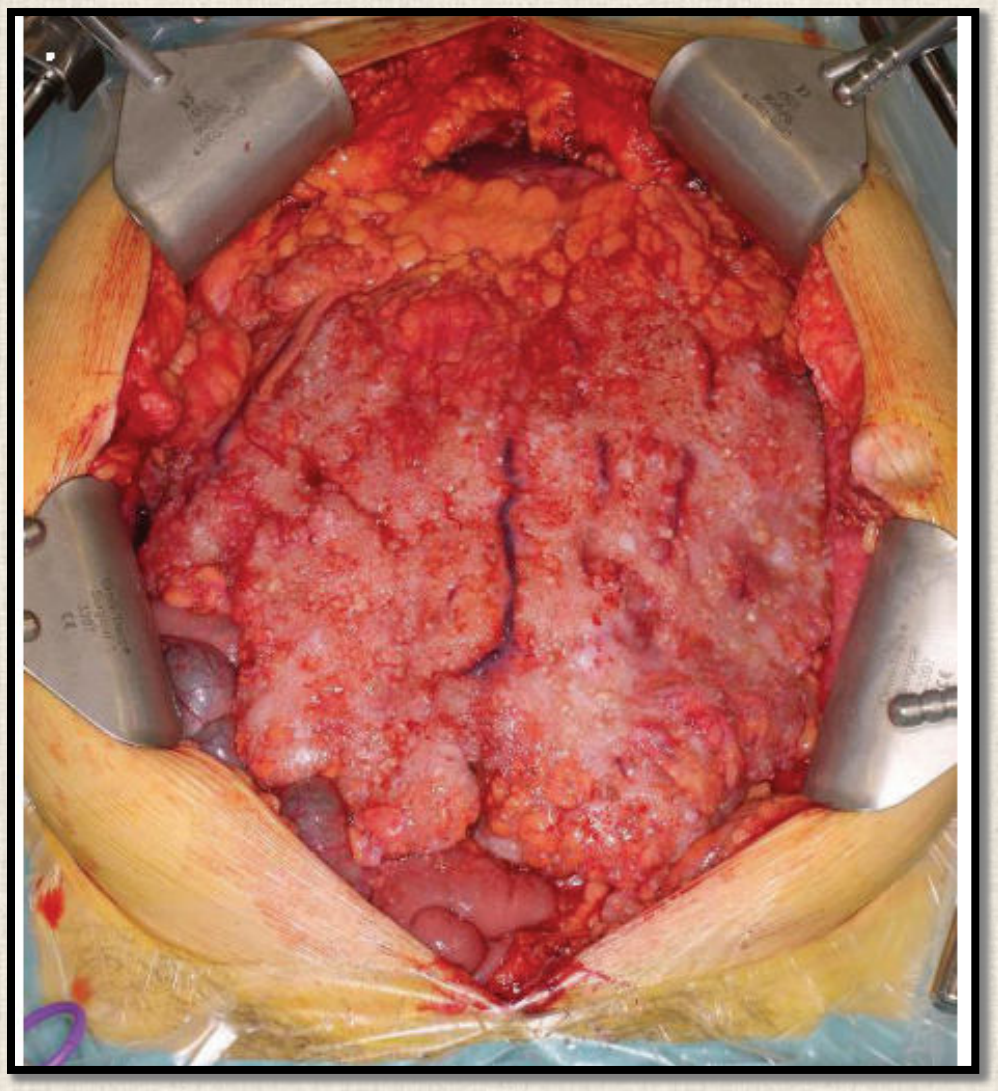
- Retention cysts
- Mucosal hyperplasia
- Cystadenomas
- Cystadenocarcinomas

A mucocele of benign etiology is adequately treated by simple appendectomy

Pseudomyxoma Peritonei

- Diffuse collections of gelatinous fluid are associated with mucinous implants on peritoneal surfaces and omentum.
- Two to three times more common in females than males.
- Recent immunocytologic and molecular studies suggest that the appendix is the site of origin for most cases of pseudomyxoma.
- Pseudomyxoma is invariably caused by neoplastic mucous-secreting cells within the peritoneum.
- Clinical presentation: abdominal pain, distension, mass, “jelly-belly”

- Tumor Markers: CA-19.9, CEA, CA-125
- Imaging:
  - CT scanning is the preferred imaging modality
  - Role of colonoscopy in the diagnosis of PMP is minimal



Intra operative photograph of patient PMP

Complete Cytoreductive Surgery is the mainstay of treatment

- All gross disease should be removed
- Appendectomy is routinely performed
- Hysterectomy with bilateral salpingo-oophorectomy is performed in women.

At surgery a variable volume of mucinous ascites is found together with tumor deposits

Pseudomyxoma is a disease that progresses slowly and in which recurrences may take years to develop or become symptomatic.

Hyperthermic intraperitoneal chemotherapy (HIPEC)

- Highly concentrated
- Heated chemotherapy treatment
- Delivered directly to the abdomen during surgery
- Intraoperative HIPEC was initiated at the Washington Hospital Center in 1992

Lymphoma

- Extremely uncommon
- Gastrointestinal tract is the most frequently involved extranodal site for non-Hodgkin's lymphoma
- Frequency of primary lymphoma of the appendix ranges from 1 to 3% of gastrointestinal lymphomas
- Usually presents as acute appendicitis

Lymphoma

- CT scan showing appendiceal diameter greater than or equal to 2.5 cm or surrounding soft-tissue thickening should prompt suspicion of an appendiceal lymphoma
- management of appendiceal lymphoma confined to the appendix is appendectomy.
- Right hemicolectomy is indicated if there is extension of tumor beyond the appendix onto the cecum or mesentery.