

SMALL BOWEL --- ADENOCARCINOMA

Dr. C. Jeske

Case presentation

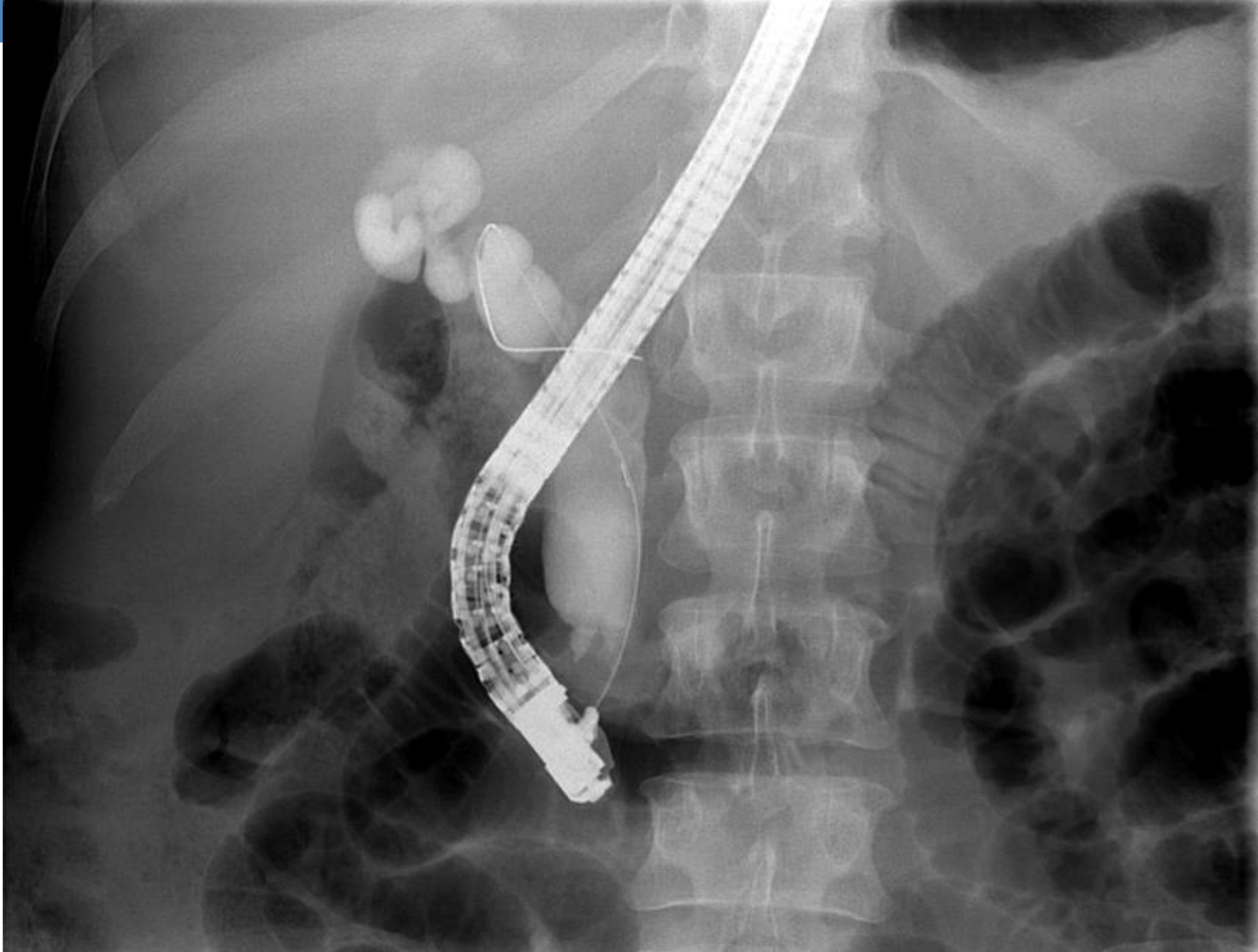
- 54 year old female.
- Presents with OJ and weight loss.
- Abdominal examination only reveals a palpable gallbladder.

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- ERCP reveals a circumferential tumour at D1/2 involving ampulla
- Stent placed
- The tumour was biopsied and confirmed moderately differentiated adenocarcinoma.



- Laparoscopy was performed to exclude peritoneal metastases.
- Pancreaticoduodenectomy was performed.
- The tumour was confined to the duodenum and there was no obvious spread.
- Awaiting final histology

Incidence

- Small bowel makes up 75% of length and 90% of mucosal surface.
- Despite this = rare cancer.
- US data = 22.7/million(2004)
- All tumours = 0.5-1.5/100000 in males and 0.2-1.0/100000 in females
- Small bowel adenocarcinoma(SBA) accounts for 40%

- Site and frequency:
 - Duodenum = 55-82%
 - Jejunum = 11-25%
 - Ileum = 7-17%
- Median age = sixth decade

Etiopathogenesis

- Environmental factors:
 - Alcohol and smoking
 - Increased risk: highest consumers of sugar, refined carbohydrates, red meat and smoked food.
 - Decreased risk: coffee, fish, fruit and vegetables.
- Lower incidence of SBA as opposed to colorectal malignancies:
 - Shorter contact time
 - Low concentration of aerophilic Gram + bacteria in SB
 - Decreased density of microbiota
 - Epithelial cells of SB – wide range of microsomal enzymes

- Carcinogenesis

- Same range of genes tested for colorectal.
- Suggests shared carcinogenesis pathway.
- However; APC mutation less often observed, MMR phenotype more frequent in SBA.
- Progress limited by small numbers and selection bias.

Genetic predisposition

- FAP
- Lynch syndrome
- Peutz-Jeghers syndrome

Other predisposing conditions

- Crohn's disease
- Coeliac disease

Clinical presentation

- Abdominal pain (43%)
- Nausea and vomiting (16%)
- Fatigue and anaemia (15%)
- Upper or lower GIT bleeding (7%)
- Jaundice (6%)
- Failure to obtain diagnostic test or misinterpretation = delays of 8-12 months.

Diagnosis

- Single center study 217 pts, diagnoses were obtained by:
 - Upper GI endoscopy (28%)
 - Surgery (26%)
 - Small bowel barium transit (22%)
 - CT scan (18%)
 - U/sound (3%)
 - Physical examination (3%)
- Diagnosis mainly obtained at advanced stages:
 - 35% synchronous metastases
 - 39% lymph-node invasion

- Sensitivity of SB barium transit and plain contrasted abdominal CT scan: 50% and 47%.
- Context of obscure bleeding after upper and lower endoscopy – SB investigation systematically done.
- Range of options.
- CT enteroclysis: sens = 85-95% and spec = 90-96%.
- Capsule endoscopy: sens = 88-95% spec = 75-95%.

Investigations after diagnosis

- Thoraco-abdomino-pelvic CT to assess distant metastases.
- Upper and lower endoscopy to look for synchronous lesions, esp. in pts with genetic predisposition.
- Baseline CEA and Ca 19-9.
- In pts with predisposing genetic disease and Crohn's – full small bowel exploration.
- Suspected Lynch syndrome according to protocol.

Prognosis

Survival according to cancer stage.

AJCC Stage	Incidence (%)	5-year OS (%)
1	4-12	50-60
2	14-30	39-55
3	19-27	10-40
4	32-46	3-5

- Lymph-node invasion = main prognostic factor
- 5-year disease free-survival = 57% 2/less nodes vs. 37% 3/> nodes.
- Significant predictors of poor OS on multivariate analysis:
 - Advanced age
 - Advanced stage
 - Ileal location
 - Recovery of <10 lymph nodes
 - Number of positive nodes

- Recurrence rate shown to be as high as 16% in one study.
- Prolonged follow-up indicated.

Treatment

- Surgery remains the only potentially curative treatment.
- Locally advanced cancer found to be irresectable at surgery = 5%.

Surgery

- Indicated for localised cancer.
- Complete resection (R0) of primary tumour + locoregional lymph node resection is mandatory.
- Context of posterior invasion – neoadjuvant treatment.
- Primary tumour resection in context of unresectable metastases is not recommended except if lesion complicates.
- Insufficient data on metastatectomy.

- **Duodenal tumours:**
Pancreaticoduodenectomy with lymph node resection.
- **Jejunal and ileal tumours:** R0 resection with lymph node resection and jejuno-jejunal or ileo-ileal anastomosis.
- **Distal ileum:** Right hemicolectomy with ligation of ileocolic artery for adequate lymph node resection.

Conclusion

- Rare cancers.
- Must maintain high index of suspicion.
- Often diagnosed at advanced stages.
- New diagnostic modalities should be used more readily.
- Better prognosis than gastric or pancreatic cancers but worse prognosis than colorectal carcinoma.