Envoi SPECIALIST PATHOLOGISTS *inside insights*

Update No.1 – 2018 *Dr Mark Bettington*

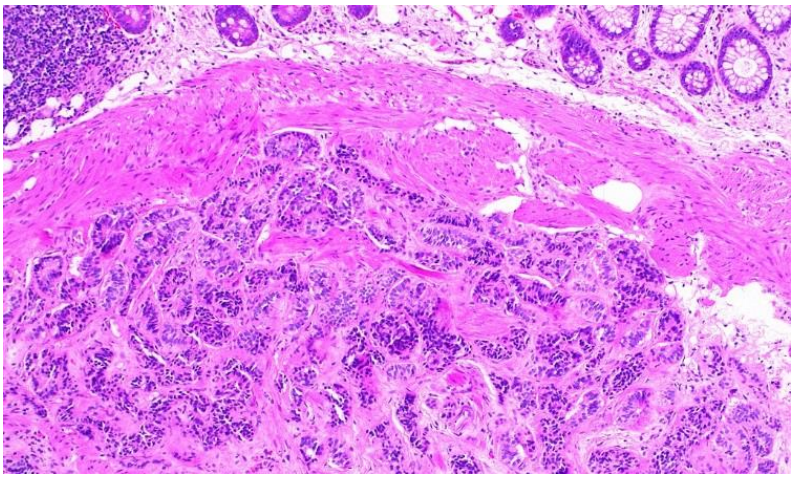
Rectal Neuroendocrine Tumours

Definition: Rectal neuroendocrine tumours (NET) ("carcinoids") are neoplasms of the hindgut and are most frequently derived from the enteroendocrine L-cells (which secrete glucagon-like peptides). They are usually small and low grade. In comparison colonic NET are often larger than 2cm and more likely to be aggressive.

Incidence: The current incidence of rectal NET is approximately 1:100 000 although this is increasing rapidly (7.8%/year).

Clinical presentation: The majority of rectal NETs are incidental lesions. At endoscopy they present as round polyps with intact overlying mucosa. They are typically yellow (due to the chromogranin granules) and can be mistaken for a lipoma.

Pathology: The histological diagnosis is typically straightforward. These lesions are composed of nests and trabeculae of bland cells with round nuclei and a characteristic "salt and pepper" chromatin pattern. The majority (88%) of rectal NETs are grade 1, thus mitoses are rare.



Staging and grading: Staging using the TNM system is dependent on size and depth of invasion. Grading is based on mitotic count and/or the proliferative index as assessed in a Ki-67 stained section.

Table 1. T staging for rectal NET

T1	Invasion lamina propria or submucosa and ≤ 2cm
	T1a <1cm
	T1b 1-2cm
T2	Invasion muscularis propria or >2cm with invasion lamina propria or submucosa
T3	Invasion into subserosa
T4	Invasion to serosa or other organs

Table 2. Grading of rectal NET

G1	Mitoses <2/10 HPF or Ki-67 index <3%
G2	Mitoses 2-20 or Ki-67 index 3-20%
G3	Mitoses >20 or Ki-67 index >20%

Important features of the pathology report: The pathology report should include the size, depth of invasion, grade, presence or absence of lymphatic or vascular invasion and the margin status.

Prognosis: In rectal NET, tumour size and lymphovascular invasion are predictors of lymph node involvement. Without lymph node metastases, survival is near 100%. 5-year survival is 54-73% for node positive disease and 15-30% with distant metastases.

Management:

- **Rectal NETs less than 10mm (T1a); Grade 1**
  - Complete removal (endoscopic or transanal)
  - No further treatment or follow-up required

- **Rectal NET 10-20mm; Grade 1 -> assess with endorectal ultrasound or MRI**
  - No invasion muscularis propria (T1b) and no lymphovascular invasion
    - Complete removal (EMR, ESD or transanal excision)
    - Endoscopy and ultrasound or MRI at 6-12 months; no long term surveillance required
  - No invasion muscularis propria (T1b) with lymphovascular invasion
    - Controversial; consider radical resection (however in one study from Japan lymphovascular invasion was not an adverse feature)
  - Invasion muscularis propria (T2) or nodal metastasis (N1)
    - CT or MRI pelvis and abdomen to identify distant metastatic disease
    - Formal resection (e.g. low / ultralow anterior resection with total mesorectal excision or abdominoperineal resection)
    - Surveillance every 6-12 months for ten years (CT or MRI pelvis, serum chromogranin A, urinary 5-HIAA)

- **Rectal NET >20mm; Grade 1**
  - Management is similar to T2 NET above

- **Rectal NET; Grade 2**
  - Grade 2 rectal NETs are rare (6% of cases) but have been demonstrated to have a worse prognosis. However specific management guidelines are not available (presumably due to limited data). In one study five year survival for grade 2 tumours was only 48% which may favour aggressive treatment for these patients.

Further reading:

Neuroendocrine tumors of the colon and rectum. Diseases of the colon and rectum. 2017;60:1018-1021
Clinical and prognostic features of rectal neuroendocrine tumors. Neuroendocrinology 2013;98:180-187
Excellent prognosis following endoscopic resection of patients with rectal neuroendocrine tumors despite the frequent presence of lymphovascular invasion. Journal of Gastroenterology 2015;50:1184-1189