Neuroendocrine tumours of the appendix

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NETs of the appendix are slightly more common in females and have an overall incidence of 0.15–0.6 per 100 /uni00A0 000 per year. They arise in subepithelial neuroendocrine cells (Figure 76.16 and the majority (70%) are located in the appendix tip. The average age at presentation is 40–50 years and most patients are asymptomatic with early-stage disease typically found at appendicectomy for acute appendicitis. Uncommonly , patients may present with symptoms due to a mass or metastatic disease. Carcinoid syndrome is extremely rare. A diagnosis of NET is based on immunohistochemical staining for synaptophysin and chromogranin A and tumours are classified as grade 1–3 according to their proliferative capacity (determined by the Ki-67 index and mitotic rate). Treatment The treatment and prognosis of NETs of the appendix is governed by their grade, the tumour size and the extent of tumour invasion. Fully resected low-grade tumours less than 1 /uni00A0 cm in size with minimal serosal or mesoappendix invasion ustria. tologist, Vienna, A Moritz Kaposi , 1837–1902, der ma are considered fully treated by appendicectomy alone and no further treatment or follow-up is required. The optimum treatment of patients with tumours 1–2 /uni00A0 cm in size that have been fully resected is less clear as metastases may occur, albeit rarely . Current guidelines recommend a single CT or MRI of the abdomen to out rule regional or distant metastatic disease. Further surgery in the form of oncological resection of the right colon should be considered in patients with - larger tumours (>2 /uni00A0 cm), in the case of incomplete resection at appendicectomy or for higher tumour grade (2 or 3), T4 disease or vascular invasion. In these patients the risk of - regional lymph node involvement is increased; however, the potential benefit of further surgery must be weighed against the increased operative risk. No further follow-up is required when the completion hemicolectomy shows no evidence of residual disease. The presence of nodal disease or high-grade tumour mandates subsequent follow-up, typically with CT or)

Figure 76.15 A low-grade mucinous tumour of the appendix with mucinous ascites and low-volume pseudomyxoma peritonei. TABLE 76.3 Classi /f_i cation of epithelial neoplasia of the appendix. Adenoma (tubular, tubulovillous, villous) Serrated polyp Non-mucinous adenocarcinoma Mucinous neoplasm Low-grade appendiceal mucinous neoplasm High-grade appendiceal mucinous neoplasm Mucinous adenocarcinoma Adenocarcinoma with signet ring cells (<50%) Signet ring (>50%) carcinoma Adapted from Carr NJ, Cecil TD, Mohamed F et al . A consensus for classi /f_i cation and pathologic reporting of pseudomyxoma peritonei and associated appendiceal neoplasia. The results of the Peritoneal Surface Oncology Group International (PSOGI) modi /f_i ed Delphi process. Am J Surg Pathol 2016; 40 : 14–26.

MRI at 6 and 12 months and then annually . For patients with early-stage NET the prognosis is excellent with 5-year survival of close to 100%. For those with advanced disease or distant metastases 5-year survival is typically less than 25%.

Figure 76.16 (a) Cross-sectional view of the appendix with outer, pink, muscularis propria (MP) and inner paler mucosa (Muc). The lumen of the /uni00A0 appendix (L) has been compressed by an adjacent well-differentiated neuroendocrine tumour (NET). Haematoxylin and eosin stain, × 20. (b) Higher power view of synaptophysin immunohistochemical stain showing characteristic positive staining (brown) within tumour cells. /uni00A0 Synaptophysin immunohistochemistry, × 100 (courtesy of Dr J Aird, FRCPath, Dublin, Ireland).

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