Pseudomyxoma peritonei

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PMP is a rare condition typified by progressive peritoneal tumour deposits, mucinous ascites, omental cake (76.17) and ovarian involvement in females. The vast majority of cases arise as a result of perforation of a mucinous appen diceal tumour. Patients typically present with progressive and massive abdominal distension, anor exia and symptoms of bowel dysfunction. The condition is invariably fatal without intervention. Traditionally , PMP was thought to have an incidence of 1 per 1 /uni00A0 000 /uni00A0 000 per year, but it is now thought to be at least double that with recent estimates of 3.2 cases per 1 /uni00A0 000 /uni00A0 000 per year. The overall risk of developing pseudomyx oma following removal of an appendix harbouring epithelial tumour is approximately 9%, with the risk varying according to the tumour subtype and the mode of presentation, while it may be as high as 30–50% in the case of a mucinous adeno carcinoma of the appendix (Figur e 76.18). PMP is classified according to the degree of cytological atypia within the peritoneal deposits (Table 76.4) and its grading may di ff er from that of the causative primary appendiceal tumour. Elevated tumour mar kers (CEA, CA-125, CA-19-9) - - Figure - - -

(b) Figure 76.18 (a) Contrast-enhanced axial computed tomography (CT) image demonstrates a tubular cystic structure with calci /f_i cation adjacent to the caecum compatible with an abnormally distended appendix (arrow). (b) Six-year follow-up postcontrast axial CT image demonstrated a 19 × 10 × 17 cm complex cystic mass in the right lower quadrant (arrow) highly suspicious for a mucinous tumour of the appendix with extensive peritoneal involvement and pseudomyxoma peritonei (courtesy of Professor Helen Fenlon, Dublin, Ireland). TABLE 76.4 Classi /f_i cation of pseudomyxoma peritonei. Acellular mucin Low-grade mucinous carcinoma peritonei High-grade mucinous carcinoma peritonei High-grade mucinous carcinoma peritonei with signet ring cells 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.

predict a more aggressive phenotype and are associated with a worse prognosis.

P T Figure 76.19 Hyperthermic intraperitoneal chemotherapy (HIPEC) delivery following cytoreductive surgery using a closed abdomen technique. The perfusion pump (P) heats and circulates chemother apy throughout the abdominal cavity via in /f_l ow and out /f_l ow tubing (T), typically for 60–90 minutes.

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