Answer

Low-Grade Appendiceal Mucinous Neoplasm

Mucoceles have been described as benign appendiceal lesions. However, appendiceal mucoceles include any cystic, mucin-filled mass in the appendix including neoplasms.[1] Although there is no consensus on the classification of primary epithelial tumors of the appendix, they are characterized as either mucinous cystadenomas or mucinous cystadenocarcinomas. However, neither classification reflects the complex behavior of these tumors. Misdraji and Young[2] introduced the term low-grade appendiceal mucinous neoplasm (LAMN), which includes lesions that have been described as villous adenoma, cystadenoma, mucinous tumor of uncertain malignant potential, and borderline tumor of the appendix.

In a review of 107 appendiceal mucinous tumor specimens, 82% were LAMNs.[3] These tumors are most common in patients aged 60 years or older, with a female to male patient ratio of 3.9:1.[2,3] Patients may be asymptomatic or have abdominal pain similar to acute appendicitis. They can also have intestinal obstruction, intussusception, rectal bleeding, ureteral obstruction, mucin-containing hernias, or hematuria.[2,4] The gross appearance of the specimen was a nondilated appendix, with a well-encapsulated mass at the tip measuring $7.6 \times 6.0 \times 5.0$ cm with no evidence of perforation. This is unusual because 69% of appendices with LAMN are often dilated or cystic.[3]

On computed tomographic scans, LAMN can appear as a cystic mass with low attenuation, irregular wall thickening, and an absence of appendiceal inflammation.[4] However, there is no association with the thickness of the wall on computed tomographic scan and malignancy.[4] Approximately 50% of patients also have mural calcification. On magnetic resonance imaging, a LAMN appears as a cystic mass with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[5] Calcification may also be identified on T2-weighted imaging as a peripheral rim with low signal intensity.[5]

Pseudomxyoma peritonei is a rare condition with peritoneal mucinous implants and mucinous ascites. It is associated with a poor prognosis, with a 5-year survival rate of 25%.[6] Previously, pseudomxyoma peritonei automatically classified appendiceal mucinous tumors as mucinous cystadenocarcinomas. However, Misdraji and Young[2] noted that in some cases, the underlying primary tumor did not exhibit malignant histology; therefore, LAMN was deemed a more appropriate term. This reclassification emphasized that the rupture of benign neoplasms could have a significant impact on a patient's prognosis, although not as severe as patients with ruptured mucinous cystadenocarcinoma. Misdraji and colleagues[3] found that 27 patients with LAMN confined to the appendix were alive at 6 years without any evidence of disease recurrence. In contrast, the 10-year survival rate of patients with LAMN with extra-appendiceal spread was 45%.

A lack of consensus also exists about the optimal surgical management of LAMN. A formal right hemicolectomy has been recommended for all noncarcinoid appendiceal tumors.[7] However, Misdraji and Young[2] advocate that appendectomy alone is an adequate operation for LAMN, even in cases of rupture. Tumors that are located at the base of the appendix may require a right hemicolectomy to achieve negative margins.[4] In our case, the surgical margins were negative for tumor; therefore, an appendectomy was appropriate. The role of laparoscopy in removing these tumors is controversial because of the concern about spread of mucinous material in the abdomen and the possibility of seeding port sites. Despite this concern, successful resection of appendiceal mucoceles via the laparoscopic approach has been reported in a series of 8 patients with no evidence of disease at 2-year follow-up.[8] During resection of LAMN, the surgeon should meticulously search for any mucinous deposits, particularly in the right retrohepatic space, pelvis, omentum, and left paracolic space.[4] Patients with evidence of peritoneal deposits should undergo surgical debulking. Intraperitoneal chemotherapy has also been used in some patients.[9]

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