implants on peritoneal surfaces and omentum. Pseudomyxoma is two to three times more common in females than in males. Recent immunocytologic and molecular studies suggest that the appendix is the site of origin for the overwhelming majority of cases of pseudomyxoma. Pseudomyxoma is invariably caused by neoplastic mucus-secreting cells within the peritoneum. These cells may be difficult to classify as malignant because they may be sparse, widely scattered, and have a low-grade cytologic appearance. Patients with pseudomyxoma usually present with abdominal pain, distention, or a mass. Primary pseudomyxoma usually does not cause abdominal organ dysfunction. However, ureteral obstruction and obstruction of venous return can be seen. Pseudomyxoma is a disease that progresses slowly and in which recurrences may take years to develop or become symptomatic. In a series from the Mayo Clinic, 76% of patients developed recurrences within the abdomen. Lymph node metastasis and distant metastasis are uncommon.

The use of imaging before surgery is advantageous to plan surgery. CT scanning is the preferred imaging modality. At surgery, a variable volume of mucinous ascites is found together with tumor deposits involving the right hemidiaphragm, right retroperitoneal space, left paracolic gutter, ligament of Treitz, and the ovaries in women. Peritoneal surfaces of the bowel are usually free of tumor. Thorough surgical debulking is the mainstay of treatment. All gross disease and the omentum should be removed. If not done previously, appendectomy is routinely performed. Hysterectomy with bilateral salpingo-oophorectomy is performed in women. Survival is better in patients who undergo R0 or R1 resection than in patients who undergo R2 resection (visible gross disease remaining). Because 5-year survival of mucinous appendiceal neoplasms is only 30%, adjuvant intraperitoneal hyperthermic chemotherapy is advocated as a standard adjunct to radical cytoreductive surgery. Cytoreductive surgery with intraperitoneal hyperthermic chemotherapy is a long, tedious procedure with operative times of 300 to 1020 minutes reported. In addition, morbidity (38%) and mortality (6%) are high. Cytoreductive surgery with intraperitoneal hyperthermic chemotherapy is associated with a 5-year survival rate of 53% to 78%. Survival is associated with initial patient performance status.

Any recurrence should be investigated completely. Recurrences are usually treated by additional surgery. It is important to note that surgery for recurrent disease is usually difficult and is associated with an increased incidence of enterotomies, anastomotic leaks, and fistulas.

Lymphoma
Lymphoma of the appendix is extremely uncommon. The gastrointestinal tract is the most frequently involved extranodal site for non-Hodgkin’s lymphoma. Other types of appendiceal lymphoma, such as Burkitt’s lymphoma, as well as leukemia, have also been reported. Primary lymphoma of the appendix accounts for 1% to 3% of gastrointestinal lymphomas. Appendiceal lymphoma usually presents as acute appendicitis and is rarely suspected preoperatively. Findings on CT scan of an appendiceal diameter ≥2.5 cm or surrounding soft tissue thickening should prompt suspicion of an appendiceal lymphoma. The management of appendiceal lymphoma confined to the appendix is appendectomy. Right hemicolecotomy is indicated if tumor extends beyond the appendix onto the cecum or mesentery. A postoperative staging workup is indicated before initiating adjuvant therapy. Adjuvant therapy is not indicated for lymphoma confined to the appendix.

References

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