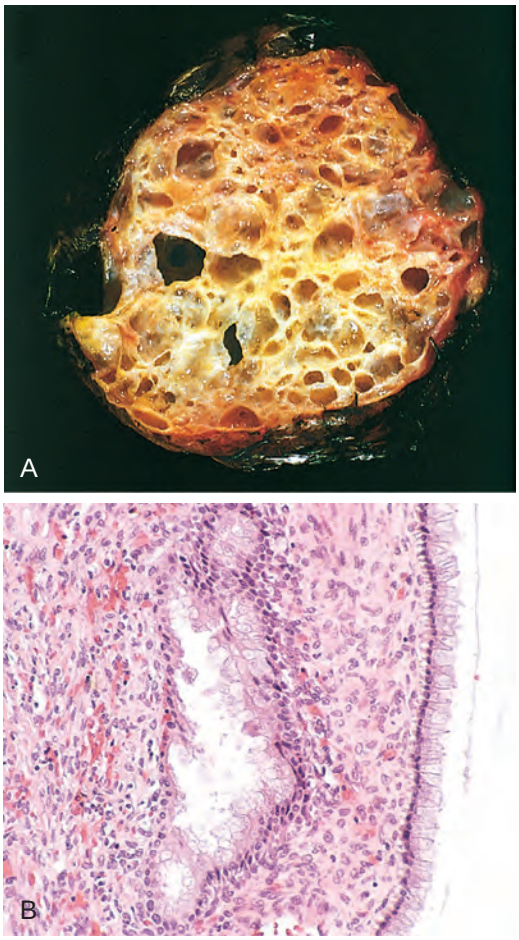

Figure 22-34 Mucinous cystadenoma **A,** Note the multicystic appearance, delicate septa, and the presence of glistening mucin within the cysts. **B,** Columnar cells lining the cysts.

A clinical condition referred to as *pseudomyxoma peritonei* is marked by extensive mucinous ascites, cystic epithelial implants on the peritoneal surfaces, adhesions, and frequent involvement of the ovaries (Fig. 22-35). Pseudomyxoma peritonei, if extensive, may result in intestinal obstruction and death. Historically, it was thought that many cases in women were due to the spread of primary ovarian mucinous neoplasms. However, recent evidence points to the source being, in almost all cases, extraovarian (usually appendiceal) (Chapter 18). Because the majority of primary mucinous ovarian tumors are unilateral, bilateral presentation of mucinous tumors always requires exclusion of a nonovarian origin.

Endometrioid Ovarian Tumors

Endometrioid carcinoma accounts for approximately 10% to 15% of all ovarian cancers. Benign endometrioid tumors, called *endometrioid adenofibromas*, and borderline endometrioid tumors also occur, but are uncommon. Endometrioid tumors are distinguished from serous and mucinous tumors by the presence of tubular glands resembling benign or malignant endometrium. Endometrioid carcinomas may arise in the setting of endometriosis and are occasionally associated with areas of borderline tumor.

Although these tumors are less common than either serous or mucinous tumors, more is known about the molecular genetic alterations associated with their development. This is due to the recent development of mouse models that closely mimic the human disease and molecular genetic overlap with endometrioid carcinomas of the endometrium. In fact, 15% to 30% of ovarian endometrioid carcinomas are accompanied by carcinoma of the endometrium, and the relatively good prognosis in such cases suggests that the two arise independently rather than by metastatic spread.

Pathogenesis. **About 15% to 20% of cases with endometrioid carcinoma coexist with endometriosis**. The peak incidence of tumors associated with endometriosis occurs a decade earlier than that of endometrioid carcinomas that are not associated with endometriosis. Molecular studies have found striking similarities to endometrial endometrioid carcinoma; shared features include relatively frequent alterations that increase PI3K/AKT pathway signaling (mutations in *PTEN*, *PIK3CA*, *ARID1A*, and *KRAS*) and mutations in mismatch DNA repair genes and *CTNNB1* (β-catenin). As mentioned earlier, mutations in *PTEN* have also been found in atypical endometriosis suggesting that it occurs early in the pathogenesis of ovarian endometrioid carcinoma, as it does in endometrioid carcinoma of the endometrium. Also similar to endometrioid carcinomas of the endometrium, *TP53* mutations are common in poorly differentiated tumors.

MORPHOLOGY

Endometrioid carcinomas typically present with solid and cystic areas of growth. Forty percent involve both ovaries, and such bilaterality usually implies extension of the neoplasm beyond the genital tract. These are low-grade tumors that reveal glandular patterns bearing a strong resemblance to those of endometrial origin. The 5-year survival rate for patients with stage I tumors is approximately 75%.

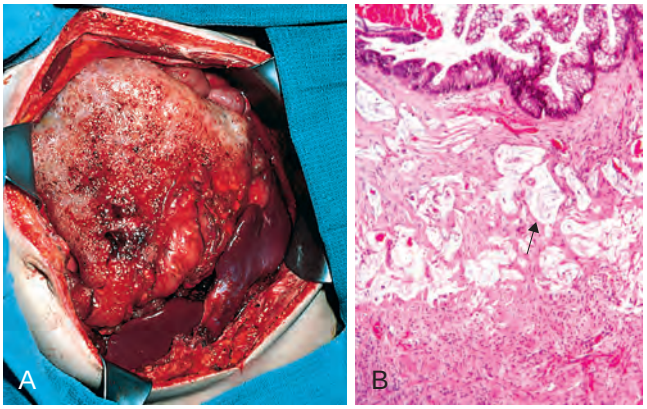

Figure 22-35 Pseudomyxoma peritonei. **A,** View at laparotomy revealing massive overgrowth of a gelatinous metastatic tumor. **B,** Histology of peritoneal implants from an appendiceal tumor, showing mucin-producing epithelium and free mucin *(arrow)*. (**A,** Courtesy Dr. Paul H. Sugarbaker, Washington Hospital Cancer Center, Washington, D.C.)