Histovathology

Small cell carcinoma of the endometrium has features resembling small cell carcinoma of the lung. The growth pattern is usually diffuse with extensive necrosis. The cells are small or of intermediate size with hyperchromatic nuclei, numerous mitotic figures and scanty cytoplasm. Immunohistochemical evidence of neuroendocrine differentiation is demonstrated in most tumors (neuron-specific enolase, chromogranin, synaptophysin, leu-7). Some tumors show an admixture with ordinary adenocarcinoma or squamous cell differentiation.

Prognosis

In contrast to the common types of endometrial carcinoma, most patients with small cell carcinoma present with advanced stage and die of widespread disease within a year. In the Norwegian series, the 5-year survival was 64%. This is probably due to a better stage distribution compared to other published series with this disease. The mean age is about 58 years.

Differential diagnosis

The differential diagnosis is with malignant lymphoma, leukemia, stromal sarcoma, embryonal rhabdomyosarcoma, primitive neuroectodermal tumors and malignant mixed mOllerian tumor.

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Mesenchymal tumors of the uterus

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This paper confines itself to discussing recent changes in opinion or understanding of uterine pure mesenchymal tumors.

Smooth muscle tumors

Mitotically active leigmyomas

It has become clearer in the past 5 years what the relative value of mitotic counts in uterine smooth muscle tumors (SMTs) is. Previous recommendations regarding mitotic counts and outcome are now known to have been too dogmatic. Leiomyomata without significant cytological atypia (none or only mild atypia) may have mitotic counts up to 20 per 10 high power field (HPF) and still pursue a benign course. Such tumors tend to occur in reproductive life and to be seen especially in the secretory phase of the cycle. Mitotically active leiomyomata should not show abnormal mitoses or coagulative necrosis. Cytologically bland tumors with more than 20 mitoses per 10 HPF are not yet sufficiently well understood for definite comment.

Lelomyomas with bizarre nuclei

When lelomyomas with bizarre nuclei show focal atypia without coagulative necrosis or abnormal mitoses, they may behave as benign tumors even with mitotic counts above 10 per HPF. In the series of Bell *etal.,* diffuse atypia and a mitotic count above 10 per 10 HPF was associated with malignant behavior. A more recent paper suggests that half of bizarre leiomyomas" show diffuse atypia and confirms that maximum mitotic rates of up to 7 per 10 HPF may be seen without malignant behavior.

Eoithelioid smooth muscle tumors

Well-defined margins, a clear cell pattern, hyalinization and the absence of necrosis are good prognostic features in epithelioid SMTs. Mitoses may be low, however, even in frank epithelloid leiomyosarcomas, with some having mitotic rates as low as 3-4 per 10 HPF. Most, but not all, malignant examples show pleomorphism, necrosis and hypercellularity and caution is required in the diagnosis of these tumors.

Myxoid smooth muscle tumors

Myxoid SMTs are uncommon but increased mitotic rates and plecmorphism etc. may be absent even in highly aggressive examples. The presence of an infiltrative margin and a size greater than 4.0 cm are indications of malignancy and all myxoid SMTs must be treated with caution. They should be differentiated from leiomyomas with hydropic degeneration and "perinodular hydropic leiomyomas.

Cellular smooth muscle tumors

Cellularity *per* se is not a worrying feature in SMTs unless accompanied by pleomorphism etc.. Cellular SMTs may, however, be very difficult to distinguish from stromal tumors. Unlike stromal tumors they tend to merge with the myometrium at their periphery and have large, thick walled blood vessels in contrast to the small arterioles typical of stromal tumors. Cellular lelomyomas may also show cleft-like spaces. Immunohistochemistry is of limited value.

Hemorrhagic cellular (aooolectic.l leiomvomas

Pregnancy, progesterone therapy and gonadotropin-releasing hormone analog therapy may all produce worrying changes in benign SMTs, including pleomorphism, infarction, increased mitoses and hemorrhage. The keys to the correct interpretation of these benign, but worrying, changes are the clinical history, lack of coagulative necrosis, well-defined tumor margins and occurrence of the changes in several tumors.

Stromal tumors

The classical differentiation of stromal tumors into stromal nodules, low-grade stromal sarcomas and high-grade stromal sarcomas has

been challenged in recent years, with many arguing that most cases of high-grade stromal sarcoma are simply undifferentiated sarcomas. The differentiation of stromal nodule from low-grade stromal sarcoma depends on demonstrating an infiltrative margin and/or vessel invasion in the latter. Cases of low-grade stromal sarcoma confined to the uterus at original presentation appear to have a very low risk of recurrence.

Recently the spectrum of differentiation patterns in low-grade stromal sarcoma has been extended. As well as sex-cord-like areas, rhabdoid and retiform differentiation have recently been described. True glands and tubules are well described and it seems likely that most examples of aggressive endometriosis represent low-grade stromal sarcoma with extensive endometriold glandular differentiation.

It is common for stromal tumors to show areas of smooth muscle differentiation. Those where more than 30% of the tumor was made up of the lesser element (sometimes called stromomyomas) have recently been the subject of a study which suggests that they should be reported as endometrial stromal nodules or low-grade stromal sarcomas with smooth muscle differentiation, as the one tumor of seven with follow-up that recurred, did so as a pure low-grade stromal sarcoma.

Sarcoma botrvoides of the cervix of young women

Several recent papers have confirmed Daya and Scully's observation that sarcoma botryoldes is usually cured by local therapy (though in the literature such local removal has usually been combined with chemotherapy). It is a tumor occurring mainly in teenagers (mean age 18). Eighty percent of cases appear free of disease at 5 years though it is not possible on histological appearances to predict those cases that will follow an aggressive course.

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