neoplasm unrelated to ESTSCLE and endometrial stromal tumors.[17,18] Interestingly, Wang et al19 described a case of UTROSCT with t(X;6)(p22.3;q23.1) and t(4;18)(q21.1;q21.3). Various known tumor-associated genes (bcl2, MALT1 and DCC at 18q21; and RAP1 at 4q21) and a gene related to the embryogenesis of gonads such as H-Y regulator gene at Xp22.3 are located at or near the translocation breakpoints. The tumor cells of sex-cordlike elements in this case showed strong and diffuse immunoreactivity for BCL2. These cytogenetic and IHC data may suggest potential molecular mechanisms of tumorigenesis for UTROSCT.[19]

In the 1976 article by Clement and Scully,[2] an origin from the endometrial stromal cells, adenomyosis, stromal myosis, endometriosis, or multipotential myometrial cells was postulated for these stromal tumors. Various studies postulated stromal, epithelial, smooth muscle, sex-cord differentiation, or pluripotent mesenchymal cell origin. The histogenesis of UTROSCT is uncertain, but endometrial stroma has been suggested. A more recent ultrastructural study on 13 cases of UTROSCT has shown that these tumors display epithelial and sex-cordlike differentiation but no smooth muscle differentiation, which supports a polyphenotypic histogenesis.[10]

Although UTROSCT is a distinct histopathologic entity, several benign and malignant neoplasms can cause a diagnostic dilemma. Some of these distinctions are of morphologic importance, and others are important from a prognostic standpoint. These include epithelioid leiomyoma, low-grade endometrial stromal sarcoma with sex-cord elements, endometrioid carcinoma with sex-cordlike features, plexiform tumorlet, vascular plexiform leiomyoma, and metastatic ovarian sex-cord stromal tumors.[11,20–25] Other conditions that may enter into the differential diagnostic consideration less frequently are adenosarcoma, carcinosarcoma, perivascular epithelioid cell tumor, and adenomatoid tumor.[26–32]

Epithelioid leiomyoma is a form of uterine leiomyoma with more than 50% round to polygonal cells. Epithelioid leiomyoma and UTROSCT show striking macroscopic resemblance. Both conditions present as a well-circumscribed, intramural mass with soft consistency, and yellow to tan cut surfaces. Immunohistochemically, these tumors are positive for epithelial and smooth muscle markers; however, epithelioid leiomyoma lacks the typical sex-cord phenotype of UTROSCT.[20] Low-grade endometrial stromal sarcoma with sex cord elements is a rare, malignant tumor of the uterine corpus. Histologically, low-grade endometrial stromal sarcoma has an infiltrative margin and diffuse growth pattern with very few scattered glands or tubules. Low-grade endometrial stromal sarcoma is typically positive for CD10 and negative for sex cord markers.[21] These tumors generally have an indolent course.[11,22] Furthermore, as described above, endometrial stromal tumors, including low-grade endometrial stromal sarcoma, endometrial stromal nodule, and ESTSCLE, show t(7;17)(p15;q21) translocation; however, no specific or significant genetic alteration, including t(7;17), is observed in UTROSCT.[17,18] Endometrioid carcinoma with sex-cordlike features is an unusual