overlying endometrium may also be involved. The maximum dimension of the tumor ranges from 2 to 15 cm (mean, 6 cm).[9] The cut surfaces are fleshy, grey-yellow to white. Necrosis and hemorrhage are unusual.[12] Microscopically, a UTROSCT displays a variety of architectural patterns, including anastomosing cords of 1 to 2 cells wide, broad trabeculae, small nests, and sertoliform or retiform tubular structures, Call-Exner–like bodies and diffuse sheets of uniform granulosa cell tumorlike areas.[2,9–11] The neoplastic cells are usually small with round to ovoid nuclei, little nuclear pleomorphism, inconspicuous nucleoli, and scant, indistinct eosinophilic cytoplasm. In rare instances, nuclear grooves may be seen. Sometimes, the neoplastic cells are spindle shaped, suggesting a biphenotypic (mixture of epithelial and mesenchymal elements) derivation for the tumor.[14] The intervening stroma shows hyalinization with a sparse lymphocytic infiltrate accompanied by foamy histiocytes, a few multinucleated giant cells, hemosiderin deposition, and/or cholesterol crystals.[15] Mitotic figures are usually not observed, and necrosis is generally absent. The presence of Charcot-Böttcher crystals in the cells of a UTROSCT suggests that some of these lesions have a true Sertoli differentiation.[16] After analyzing the plethora of IHC data on UTROSCT, Irving et al[3] concluded that the most common and reliable IHC markers for UTROSCT are calretinin, inhibin, CD99, and Melan-A and that immunoreactivity for calretinin and for at least one of the other 3 sex cord markers in that immunopanel is highly suggestive of UTROSCT. Leval et al[6] found that UTROSCT has a diverse IHC profile with shared expression of sex cord, epithelial, and smooth muscle markers. Sex cord markers, such as calretinin (positive in up to 95.8% of cases), inhibin positive in up to 48% of cases), CD99, WT1 (positive in up to 83.3% of cases), and Melan-A (positive in up to 85% of cases); epithelial markers, such as pancytokeratin (positive in up to 50% of cases) and epithelial membrane antigen (positive in up to 34.4% of cases); and smooth muscle markers, such as smooth muscle actin (positive in up to 40.8% of cases), desmin (positive in up to 25.3% of cases), and histone deacetylase 8 (positive in up to 66.6% of cases), are expressed along with few miscellaneous markers, such as CD10 (positive in up to 60% of cases), estrogen receptor, progesterone receptor, S100 (positive in up to 18.2% of cases), and CD117 (positive in up to 33.3% of cases). The IHC profile of these tumors appears to be intermediate between that of ESTSCLE (which typically show less sex cord marker expression) and ovarian sex cord stromal tumors of the ovary (which show marked expression of sex cord markers).[6]

The UTROSCTs have diverse findings under the electron microscope, including both epithelial (desmosome-like junctions, tonofilaments, lumina formation, and microvilli) and sex-cordlike features (nuclear indentation, abundant intracellular filaments, sparse to moderate rough endoplasmic reticulum [granulosa cells], and abundant intracytoplasmic lipid [Sertoli]), along with aggregates of perinuclear filaments with concordant IHC positivity for markers of epithelial and sex cord differentiation.[10] These findings indicate that UTROSCTs are polyphenotypic neoplasms at the ultrastructural level and show evidence of variable sex-cordlike differentiation. Endometrial stromal tumors and their variants, including ESTSCLE, show t(7;17)(p15;q21) translocation, resulting in the fusion of 2 novel genes, JAZF1 and JJAZ1; however, no specific or significant genetic alteration, including t(7;17), is observed in UTROSCT. Therefore, UTROSCT most likely represents a distinct