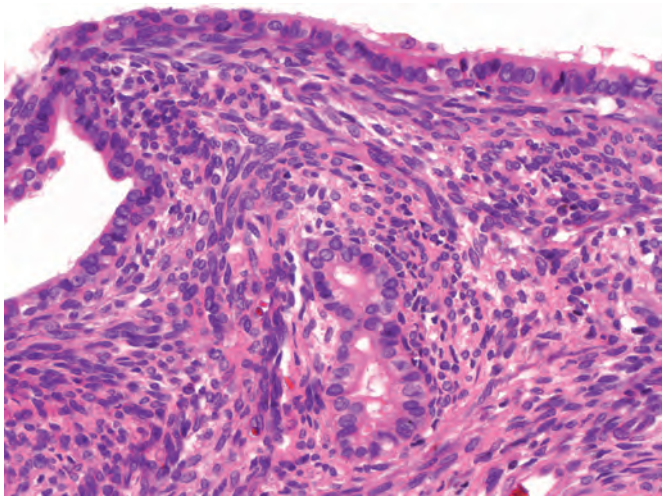

Figure 26-54 Synovial sarcoma revealing the classic biphasic spindle cell and glandlike histologic appearance.

-*SSX2*, or -*SSX4* fusion genes that encode chimeric transcription factors.

MORPHOLOGY

Synovial sarcomas are morphologically monophasic or biphasic. Monophasic synovial sarcoma consists of uniform spindle cells with scant cytoplasm and dense chromatin growing in short, tightly packed fascicles. Many tumors historically classified as fibrosarcoma likely would be classified as synovial sarcoma today. The tumors may calcify. The biphasic type contains, in addition to the spindle cell component above, gland-like structures composed of cuboidal to columnar epithelioid cells (Fig. 26-54). Immunohistochemistry is helpful in identifying these tumors, since the tumor cells, especially in the biphasic type, are positive for epithelial markers (e.g., keratins), differentiating them from most other sarcomas.

Synovial sarcomas are treated aggressively with limb-sparing surgery and frequently chemotherapy. The 5-year survival varies from 25% to 62%, related to stage and patient age. Common sites of metastases are the lung and occasionally the regional lymph nodes.

Undifferentiated Pleomorphic Sarcoma

Undifferentiated pleomorphic sarcoma (UPS) includes malignant mesenchymal tumors with high-grade, pleomorphic cells that cannot be classified into another category by a combination of histomorphology, immunophenotype, ultrastructure or molecular genetics. Despite advances in molecular characterization of sarcomas, UPS represents the largest category of adult sarcomas. Most arise in the deep soft tissues of the extremity, especially the thigh of middle aged or older adults. The diagnosis of *malignant fibrous histiocytoma* (MFH), sometimes used interchangeably with UPS, has fallen out of usage because (1) the category included both undifferentiated tumors and others that were reclassified with immunohistochemistry or molecular methods, and (2) no consensus exists for the morphologic definition of fibrohistiocytic. Not surprisingly, reproducible genetic changes are not typical of UPS. Most tumors are aneuploid with multiple structural and numerical chromosomal changes.

MORPHOLOGY

UPS are usually large, grey-white fleshy masses and can grow quite large (10 to 20 cm) depending on the anatomic compartment. Necrosis and hemorrhage are common. They consist of sheets of large, anaplastic spindled to polygonal cells with hyperchromatic irregular, sometimes bizarre nuclei (Fig. 26-55). Mitotic figures, including atypical non-symmetric forms, are abundant as is coagulative necrosis. By definition, tumor cells lack differentiation along recognized lineages.

UPS are aggressive malignancies that are treated with surgery and adjuvant chemotherapy and/or radiation. The prognosis is generally poor, with metastases arising in 30% to 50% of cases.

KEY CONCEPTS

Soft Tissue Tumors

- The category of soft tissue neoplasia describes tumors that do not fall into categories of epithelial, skeletal, central nervous system, hematopoietic or lymphoid tissues. A sarcoma is a malignant mesenchymal tumor.
- Although all soft tissue tumors probably arise from pluripotent mesenchymal stem cells, rather than mature cells, tumors can be classified into:
  - Tumors that recapitulate a mature mesenchymal tissue (e.g., skeletal muscle) can be further subdivided into benign and malignant forms.
  - Tumors composed of cells for which there is no normal counterpart (e.g., synovial sarcoma, undifferentiated pleomorphic sarcoma)
- Sarcomas with simple karyotypes demonstrate reproducible, chromosomal and molecular abnormalities which contribute to pathogenesis and are sufficiently specific to have diagnostic utility.
- Most adult sarcomas have complex karyotypes, tend to be pleomorphic and genetically heterogeneous with a poor prognosis.

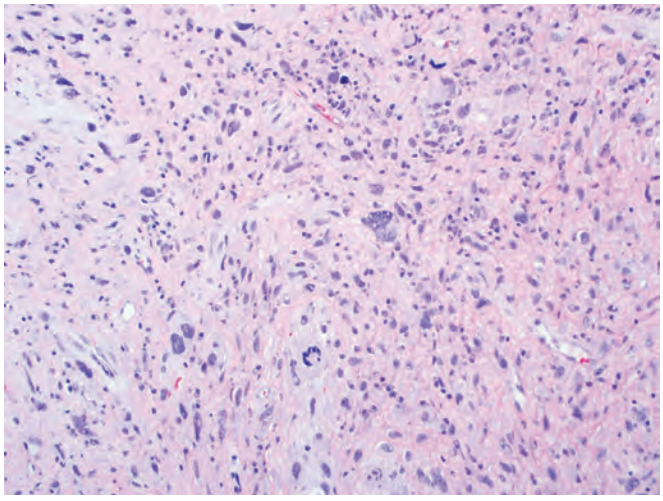

Figure 26-55 Undifferentiated pleomorphic sarcoma revealing anaplastic spindled to polygonal cells.