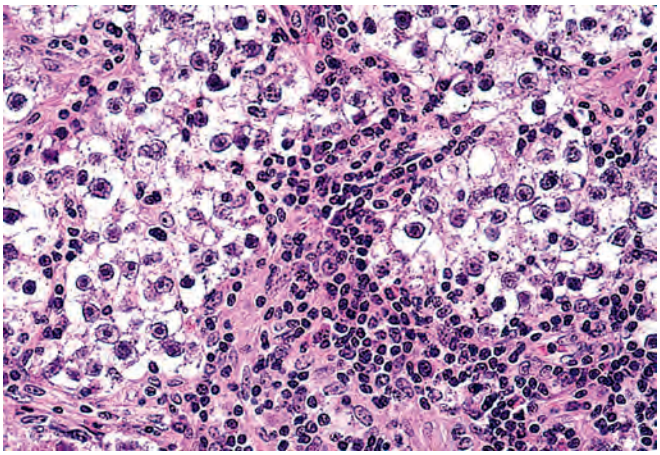

Figure 22-41 Dysgerminoma showing polyhedral tumor cells with round nuclei and adjacent inflammation.

markers and, in the case of KIT, may also serve as a therapeutic target.

MORPHOLOGY

Most dysgerminomas (80% to 90%) are unilateral tumors ranging in size from barely visible nodules to masses that virtually fill the entire abdomen. On cut surface they have a solid yellow-white to gray-pink appearance and are often soft and fleshy. Like seminoma, it is composed of large vesicular cells having a clear cytoplasm, well-defined cell boundaries, and centrally placed regular nuclei. The tumor cells grow in sheets or cords separated by scant fibrous stroma (Fig. 22-41), which is infiltrated by mature lymphocytes and may contain occasional granulomas. On occasion, small nodules of dysgerminoma are encountered in the wall of an otherwise benign cystic teratoma; conversely, a predominantly dysgerminomatous tumor may contain a small cystic teratoma.

All dysgerminomas are malignant, but the degree of histologic atypia is variable, and only about one third are aggressive. A unilateral tumor that has not broken through the capsule or spread outside the ovary has an excellent prognosis (up to 96% cure rate) after simple salpingo-oophorectomy. These neoplasms are responsive to chemotherapy, and even those that have extended beyond the ovary can often be cured. Overall survival exceeds 80%.

Yolk Sac Tumor

Though rare, yolk sac tumor (also known as *endodermal sinus tumor*) still ranks as the second most common malignant tumor of germ cell origin. It is thought that to be derived from malignant germ cells that are differentiating along the extraembryonic yolk sac lineage (Fig. 22-37). Similar to the normal yolk sac, the tumor cells elaborate *α-fetoprotein*. Its characteristic histologic feature is a glomerulus-like structure composed of a central blood vessel enveloped by tumor cells within a space that is also lined by tumor cells (*Schiller-Duval body*) (Fig. 22-42). Conspicuous intracellular and extracellular hyaline droplets are present in all tumors, and some of these stain for α-fetoprotein by immunoperoxidase techniques.

Most patients are children or young women presenting with abdominal pain and a rapidly growing pelvic mass that usually appears to involve a single ovary. With combination chemotherapy, there is greater than 80% survival independent of disease stage.

Choriocarcinoma

More commonly of placental origin, choriocarcinoma, like the yolk sac tumor, is an example of extraembryonic differentiation of malignant germ cells. It is generally held that a germ cell origin can be confirmed only in prepubertal females, because after this age an origin from an ovarian ectopic pregnancy cannot be excluded.

Most ovarian choriocarcinomas exist in combination with other germ cell tumors, and pure choriocarcinoma is extremely rare. They are histologically identical to the more common placental lesions (described later). The ovarian tumors are aggressive and have usually metastasized hematogenously to the lungs, liver, bone, and other sites by the time of diagnosis. Like all choriocarcinomas they elaborate high levels of *chorionic gonadotropins*, which may be helpful in establishing the diagnosis or detecting recurrences. In contrast to choriocarcinomas arising in placental tissue, those arising in the ovary are generally unresponsive to chemotherapy and are often fatal.

Other Germ Cell Tumors

These include (1) embryonal carcinoma, a highly malignant tumor of primitive embryonal elements that is histologically similar to embryonal carcinoma arising in the testes (Chapter 21); (2) polyembryoma, a malignant tumor containing so-called embryoid bodies; and (3) mixed germ cell tumors containing various combinations of dysgerminoma, teratoma, yolk sac tumor, and choriocarcinoma.

KEY CONCEPTS

Germ Cell Tumors

- Germ cell tumors constitute 15% to 20% of ovarian tumors.
- The majority are mature cystic teratomas (dermoid cysts) in women of reproductive age.

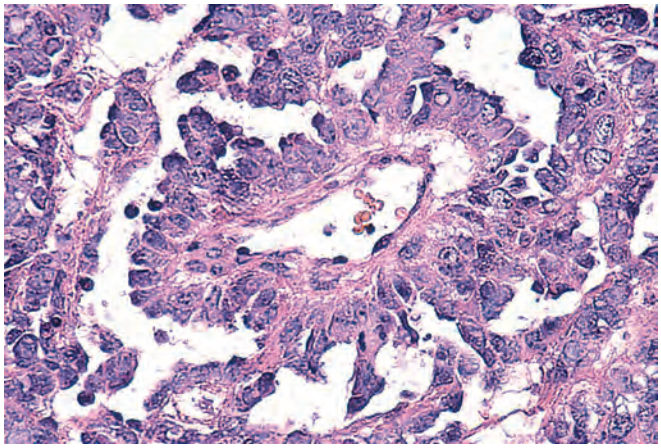

Figure 22-42 A Schiller-Duval body in yolk sac carcinoma.