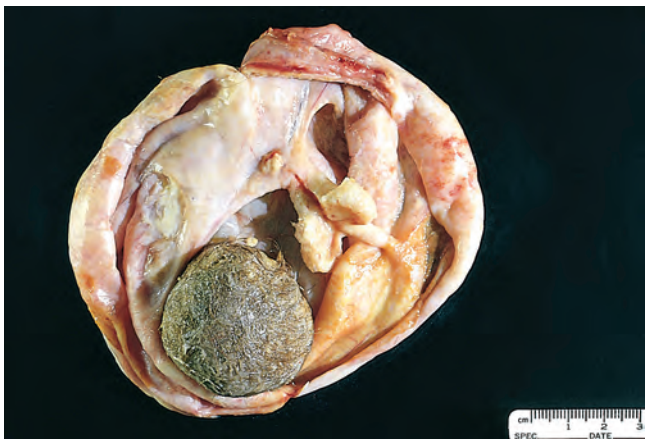

Figure 22-38 Opened mature cystic teratoma (dermoid cyst) of the ovary. Hair *(bottom)* and a mixture of tissues are evident.

The origin of teratomas has been a matter of fascination for centuries. Some common beliefs blamed witches, nightmares, or adultery with the devil. The karyotype of almost all benign ovarian teratomas is 46,XX. Genetic analyses indicate that the majority of teratomas arise from an ovum after the first meiotic division, while a minority arises before the first division.

Monodermal or Specialized Teratomas. The specialized teratomas are a remarkable, rare group of tumors, the most common of which are *struma ovarii* and *carcinoid*. They are always unilateral, although a contralateral teratoma may be present. Struma ovarii is composed entirely of mature thyroid tissue, which may be functional and cause hyperthyroidism. The ovarian carcinoid, which presumably arises from intestinal tissue found in teratomas, may also be functional; particularly if large (>7 cm), they can produce sufficient 5-hydroxytryptamine to cause the carcinoid syndrome even in the absence of hepatic metastases, since ovarian veins are directly connected to systemic circulation. Primary ovarian carcinoid must be distinguished from metastatic intestinal carcinoid, which is virtually always bilateral. Even rarer is the strumal carcinoid, a combination of struma ovarii and carcinoid in the same ovary. Only about 2% of carcinoids in teratomas metastasize.

Immature Malignant Teratomas. These are rare tumors that differ from benign teratomas in that the component tissues resemble embryonal and immature fetal tissue. The tumor is found chiefly in prepubertal adolescents and young women, the mean age being 18 years.

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The tumors are bulky and have a smooth external surface and tend to be solid on sectioning. Hair, sebaceous material, cartilage, bone, and calcification may be present, along with areas of necrosis and hemorrhage. On microscopic examination there are varying amounts of immature neuroepithelium, cartilage, bone, muscle, and other elements. An important risk for subsequent extraovarian spread is the histologic grade of tumor (I through III), which is based on the proportion of tissue containing immature neuroepithelium (Fig. 22-40).

Immature teratomas grow rapidly, frequently penetrate the capsule, and spread either locally or distantly. Stage I tumors, however, particularly those with low-grade (grade 1) histology, have an excellent prognosis. Higher-grade tumors confined to the ovary are generally treated with prophylactic chemotherapy. Most recurrences develop in the first 2 years, and absence of disease beyond this period carries an excellent chance of cure.

Dysgerminoma

Dysgerminoma is the ovarian counterpart of testicular seminoma. Dysgerminomas account for about 2% of ovarian cancers and roughly 50% of malignant ovarian germ cell tumors. They may occur in childhood, but 75% occur in the second and third decades. Some occur in patients with gonadal dysgenesis, including pseudohermaphroditism. Most of these tumors have no endocrine function. A few produce elevated levels of chorionic gonadotropin, a finding that correlates with the presence of syncytiotrophoblastic giant cells. Like seminomas, dysgerminomas express OCT-3, OCT4, and NANOG. These transcription factors are implicated in maintenance of pluripotency. They also express the receptor tyrosine kinase KIT and approximately one third have activating mutations in the *KIT* gene. These proteins are useful diagnostic

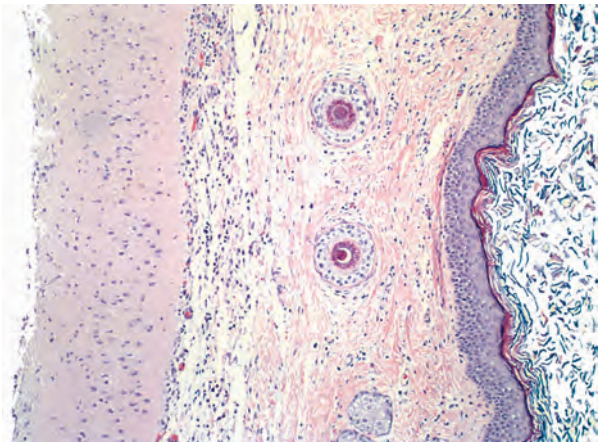

Figure 22-39 Benign cystic teratoma. Low-power view of skin *(right edge)*, beneath which there is brain tissue *(left edge)*.

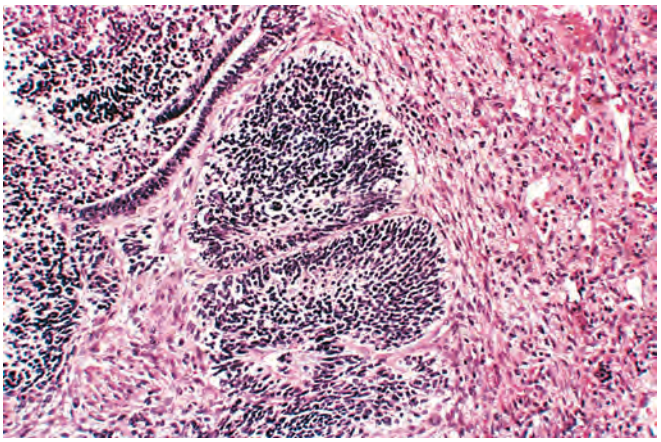

Figure 22-40 Immature teratoma of the ovary illustrating primitive neuroepithelium.