

Embryonal rhabdomyosarcoma of the ear

Nathan B. Sautter, MD

Lester D.R. Thompson, MD

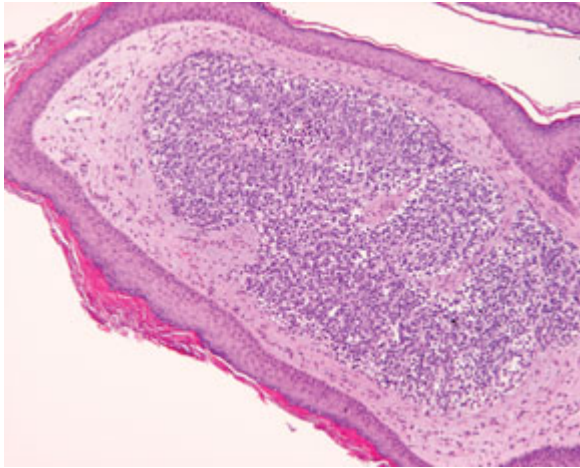


Figure 1. An intact metaplastic squamous mucosa overlies a compact neoplastic proliferation of rhabdomyoblasts with a small area of necrosis (H&E, intermediate magnification).

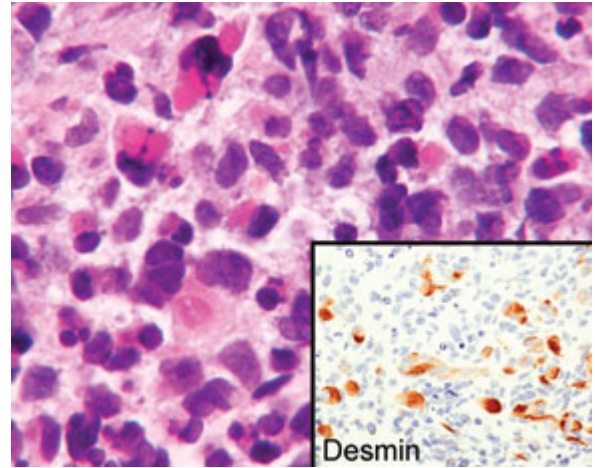


Figure 2. The cells of embryonal rhabdomyosarcoma are round to spindle shaped with eccentrically placed eosinophilic cytoplasm (H&E, high magnification). The nuclei are pleomorphic. Inset: Cells demonstrate cytoplasmic immunoreactivity with desmin, helping confirm the diagnosis of rhabdomyosarcoma.

Rhabdomyosarcoma is the most common soft-tissue malignancy in the pediatric population. It is generally classified into embryonal, alveolar, pleomorphic, and mixed histologic subtypes. Embryonal rhabdomyosarcoma is the most common histologic variant seen in childhood; a large proportion of them arise in the head and neck—most commonly in the orbit, the nasopharynx, and the ear.

Rhabdomyosarcoma of the middle ear and mastoid is the most common malignant aural neoplasm in the pediatric population, although it accounts for less than 10% of all cases of head and neck rhabdomyosarcoma. Symptoms (unilateral refractory otitis media, serosanguineous discharge, otalgia, hearing loss, and neurologic symptoms) are generally nonspecific, which can lead to a delay in diagnosis. A polypoid mass is frequently seen on examination, and it may be mistaken for an aural polyp. Because rhabdomyosarcoma of the ear is easily misdiagnosed, advanced disease with meningeal involvement is common at the time of diagnosis.

Microscopically, the surface epithelium is usually intact and separated from the neoplastic proliferation (figure 1). Embryonal rhabdomyosarcoma is made up of round to spindled cells. The appearance of the round cells is similar to that of lymphocytes, with hyperchromatic, irregular nuclei surrounded by scant, elongated eosinophilic cytoplasm. These cells imperceptibly blend with spindled cells, which are characterized by a spindled morphology, eosinophilic cytoplasm, and an elongated central hyperchromatic nucleus. Cross striations are difficult to identify, but they are rarely present. These cells are surrounded by a loose myxoid to dense collagenous stroma. Periodic acid-Schiff and a variety of immunohistochemical stains (smooth-muscle actin, desmin [figure 2], MyoD1, and myogenin) may help confirm the diagnosis.

Treatment is based on staging criteria developed by those involved in the Intergroup Rhabdomyosarcoma Studies. The multimodal approach includes wide local excision, radiation therapy, and multiagent chemotherapy. Prognosis for ear rhabdomyosarcoma is good (70% cure rates) and is related to the patient's age, tumor stage, and histologic subtype.

Suggested reading

Hawkins DS, Anderson JR, Paidas CN, et al. Improved outcome for patients with middle ear rhabdomyosarcoma: A children's oncology group study. *J Clin Oncol* 2001;19:3073-9.

Maurer HM, Beltangady M, Gehan EA, et al. The Intergroup Rhabdomyosarcoma Study-I. A final report. *Cancer* 1988;61:209-20.