

Embryonal rhabdomyosarcoma of the ear

Nathan B. Sautter, MD
Lester D.R. Thompson, MD

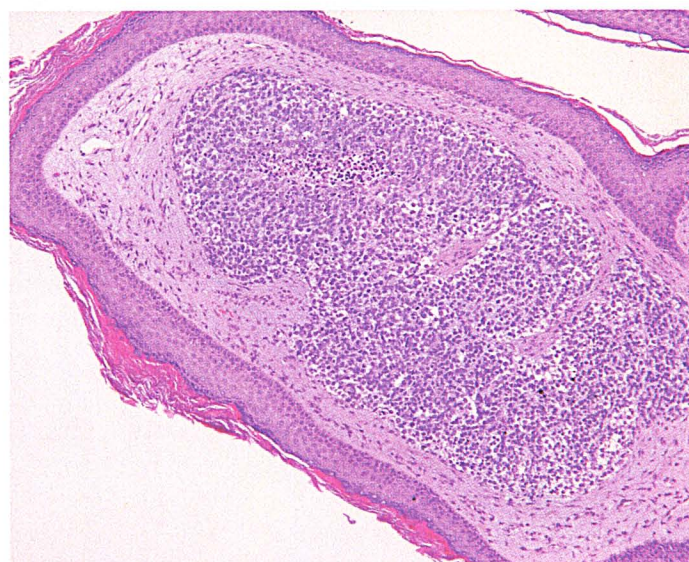


Figure 1. An intact metaplastic squamous mucosa overlies a compact neoplastic proliferation of rhabdomyoblasts with a small area of necrosis (H&E, intermediate magnification).

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 hyper-

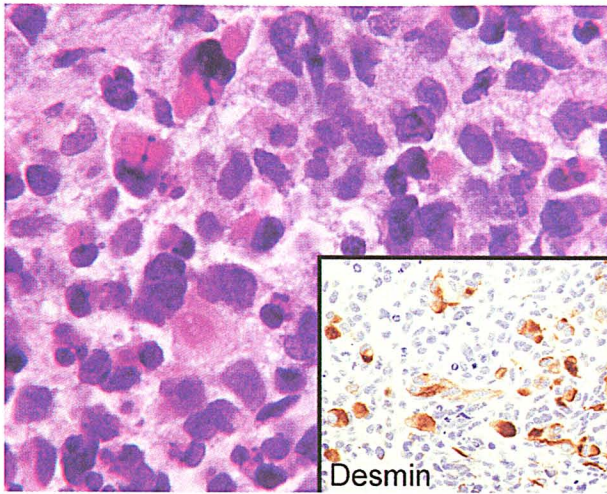


Figure 2. The cells of embryonal rhabdomyosarcoma are round to spindle shaped with eccentrically placed eosinophilic cytoplasm (H&E, high magnification). The nuclei are pleomorphic. Inset: Cells demonstrate cytoplasmic immunoreactivity with desmin, helping confirm the diagnosis of rhabdomyosarcoma.

chromatic 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.



I found more.

"CompHealth really helped me find the dream job I've been looking for."

Dr. Genise Fraiman, Internal Medicine

Call us today and put one of our consultants to work for you.

ILLINOIS—BE YOUR OWN BOSS! Solo opportunity with shared 1:3 call. Enjoy 100% of the local market and the freedom to develop any subspecialty. First-year income guarantee plus sign-on bonus, paid overhead and hospital-sponsored marketing program. High income potential and low managed care. Beautiful Chicago suburb with excellent schools. Contact Ron Bansak, 800-365-8900, ext. 248; rbansak@comphealth.com. Ref. #613041 EOE/AA

CALIFORNIA—BE BUSY FROM DAY ONE. Seeking an established ENT to join this hospital and be set up in a booming practice. Your success is virtually guaranteed, as you are assured a steady stream of patients. Practice out of a clinic located within walking distance to the hospital. Enjoy a very competitive income guarantee and full benefits. This wonderful community features excellent schools,

golfing, boating, fishing, hiking, mountain climbing and skiing, as well as easy access to San Francisco and the mountains. Contact Jodi Lloyd, 800-328-3666, ext. 7540; jlloyd@comphealth.com. Ref. #91784 EOE/AA

WEST VIRGINIA—PARTNERSHIP FAST TRACK. Join this renowned single-specialty group with an academic affiliation and perform general ENT procedures with the option to develop any subspecialty. Enjoy quick partnership, unlimited earning potential and a comprehensive benefits package. This great college town is located in the tri-state region of Ohio, West Virginia and Kentucky. Contact Robert Longmuir, 800-365-8900, ext. 214; rlongmuir@comphealth.com. Ref. #612815 EOE/AA

www.comphealth.com/entjnl. Call us at 800.328.3666 or 800.365.8900

WEST VIRGINIA—\$50K SIGN ON BONUS! Either join an established single-specialty group or go solo with an income guarantee. Both opportunities offer a fantastic earning potential and 100% support from the local hospital, which is one of the largest and most profitable in the state. Your success is assured, as the service population is huge. Practice general ENT with some facial plastics and share a call of 1:3 with area physicians. This breathtaking location features beautiful mountains and lakes, and offers easy access to surrounding cities via a major interstate highway. Contact Maureen Prunotto, 800-365-8900, ext. 239; mprunotto@comphealth.com. Ref. #611123 EOE/AA

