Temporal bone schwannoma

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A schwannoma (acoustic neuroma or neurilemmoma) is a globular, firm, tan-yellow, solid to cystic neoplasm of nerve sheath (Schwann) cells. Schwannomas are the most common neoplasms of the ear and temporal bone. The vast majority arise at the cerebropontine angle, and 95% are unilateral and sporadic. Bilateral schwannomas and those that arise in young patients are highly associated with neurofibromatosis type 2 (NF2). Schwannomas affect men and women equally, usually in the fifth or sixth decade of life except in patients with NF2, who generally present at a younger age.

Schwannomas are benign. The usual clinical manifestations are progressive, unilateral, sensorineural hearing loss and tinnitus. These symptoms are occasionally accompanied by headache, vertigo, facial pain, and facial weakness. Imaging usually demonstrates that the internal auditory canal has been widened by a mass that is isodense to the cerebellum. These tumors arise from and are attached to the vestibular division of the VIIIth cranial nerve.

Schwannomas are made up of cellular Antoni A areas with Verocay bodies (figure, A) and myxoid, hypocellular Antoni B areas (figure, B). The cells are fusiform with elongated fibrillary cytoplasm, and their buckled to spindled nuclei exhibit little atypia. Nuclear palisading is common (figure, C). Small to medium-size vessels may exhibit perivascular hyalinization. Immunohistochemistry for S-100 protein will decorate the neoplastic cells, although usually unnecessary for diagnosis. Given the unique distribution of the tumor, the only common lesions considered in the differential diagnosis are meningioma and neurofibroma.

The standard therapy is surgical removal via a number of different approaches (translabyrinthine, suboccipital, or middle cranial fossa) or by stereotactic gamma knife surgery. The 5-year survival rate is greater than 90%. The risk of recurrence is very low.

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