Congenital Cholesteatoma of the Mastoid Temporal Bone

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Purpose
Demonstrate the characteristic CT and MR findings of congenital mastoid cholesteatoma.

Materials & Methods
Clinical findings and preoperative images of three patients with congenital mastoid cholesteatoma were reviewed retrospectively. All patients underwent CT, two patients also underwent contrast-enhanced MR imaging, and one patient also had diffusion-weighted imaging and Fluid Attenuated Inversion Recovery (FLAIR).

Results
The patients presented with clinical symptoms of pain or an enlarging mass in the soft tissues overlying the mastoid region. In all patients the lesion appeared on CT as a well circumscribed nonenhancing mass with near CSF attenuation and benign erosion of the adjacent bony mastoid. In the two patients who underwent MR imaging the lesion presented as a well circumscribed mass paralleling CSF intensity on T1- and T2-weighted images with absent or thin rim-like enhancement. The lesion in the single patient who underwent additional MR sequences showed high signal (restricted diffusion) on diffusion-weighted imaging and high signal (absence of CSF signal suppression) on FLAIR.

Conclusion
Clinically mastoid cholesteatoma differs from its more common counterparts in the middle ear or cerebellopontine angle. Instead of presenting early secondary to involvement of critical structures such as the ossicles or the eighth cranial nerve, congenital cholesteatoma of the mastoid is likely to attain large size before causing symptoms and present with either pain and/or a palpable soft tissue mass following erosion through the mastoid floor. However, CT and MR characteristics in congenital mastoid cholesteatoma are identical to those found in the same tumors occurring in the middle ear and the cerebellopontine angle. On CT scans congenital mastoid cholesteatoma appears as a well circumscribed nonenhancing mass with near CSF attenuation and benign erosion of the adjacent bony mastoid. On MR imaging the lesion presents as a well circumscribed mass with absent or thin rim-like enhancement paralleling CSF intensity on T1- and T2-weighted images, high signal on FLAIR, and high signal on diffusion-weighted imaging.