Inner ear imaging: more than "rule out acoustic".

P A Hudgins

AJNR Am J Neuroradiol 1998, 19 (10) 1807-1808
http://www.ajnr.org/content/19/10/1807.citation

Inner Ear Imaging: More than "Rule Out Acoustic"

In this issue of the *American Journal of Neuroradiology* Melhem et al (page 1819) describe an MR technique to determine inner ear volumetric measurements; specifically, the total perilymphatic and endolymphatic fluid in healthy volunteers. The range of volumetric measurements in the population was large (from 172.3 to 272.6 mm^3), and the range between left and right in the same volunteers was greater than would be expected (-36.7 to 72.4 mm^3). The semicircular canals and cochlea are phylogenetically primitive and don't vary, so the wide variation in volume may be due to partial volume averaging. This suggests that, at this point, the technique may over- or underestimate volumes, but with further development we may be able to determine precise fluid volumes of the inner ear labyrinth. Indeed, this shift from gross morphologic inner ear imaging to the detection of subtle intralabyrinthine volume differences is an extraordinary development, with important applications in the diagnosis and treatment of complex inner ear disorders.

As the authors suggest, the initial application for this technique may be in understanding and diagnosing congenital labyrinthine disorders. Because the temporal bone CT scan may appear normal in the setting of congenital sensorineural hearing loss associated with a dysmorphic inner ear, otolaryngologists, neurootologists, and head and neck radiologists have investigated the use of high-resolution MR imaging. In one recent series, a patient with sensorineural hearing loss had a normal temporal bone CT scan although the MR findings were abnormal, with subtle cochlear dysplasia (1). The next logical question is obvious: Are there congenital or degenerative cochlear processes that do not affect the bony architecture (CT) and that cannot be detected on high-resolution MR images but that produce an abnormal labyrinthine volume? Congenital labyrinthine dysplasia, possibly even with a normal CT appearance, would be expected to have abnormal peri- and endolymphatic fluid volumes. Many degenerative cochlear processes (eg, postmeningitic sensorineural hearing loss) do not initially affect the bony labyrinth, but volumetric analysis of the peri- and endolymphatic fluid may show a decrease prior to the classic CT changes of labyrinthine ossificans.

Unfortunately, the common labyrinthine disorders, primarily Meniere disease (also known as endolymphatic hydrops) and perilymphatic fistula, should have normal total peri- and endolymphatic volumes, and would not be detectable with the described volumetric estimation technique. Both labyrinthine disorders, however, most likely result in a change in endolymphatic to perilymphatic fluid ratio. Our clinical colleagues, therefore, are pushing for even more precise and higher resolution imaging, as it is the separate perilymphatic and endolymphatic volumes they are most interested in—not the total labyrinthine volume. In Meniere disease, characterized by fluctuating sensorineural hearing loss, tinnitus, and vertigo, there is no imaging standard of reference. Temporal bone CT and conventional MR imaging studies are normal, except for small or nonvisualized endolymphatic ducts and sacs, a finding seen in only a subset of patients (2). Recent work in animals, however, has shown that high-resolution MR imaging allowed 3D reconstruction of each of the three labyrinthine canals: the scala tympani and scala vestibuli, both containing perilymphatic fluid, and the scala media, containing endolymphatic fluid. In animals with Meniere disease, the scala media (containing endolymphatic fluid) was enlarged compared with that of healthy control animals (3).

Perilymphatic fluid, an ultrafiltrate of CSF, is similar in ion composition to CSF. Patients with perilymphatic fistula present with progressive or fluctuating sensorineural hearing loss, sometimes associated with vertigo. Leaks of perilymphatic fluid may be small and intermittent, and no reliable preoperative test is available for diagnosis. Perilymphatic fistula, therefore, has no strict objective diagnostic criteria, and intraoperative visualization of a leak at the oval or round window remains the reference standard for diagnosis. MR imaging shows great promise in the diagnosis of perilymphatic fistula. Mark et al (4) reported a small series of patients with surgically confirmed perilymphatic fistula who had cochlear enhancement on contrast-enhanced MR images. Intrathecal contrast administration in animals with surgically created perilymphatic fistulas produced pooling of extremely small amounts of contrast agent in the middle ear or mastoids (5). Another application of the volumetric technique might be to establish an abnormal perilymphatic to endolymphatic fluid ratio in patients with suspected perilymphatic fistula, once the scala media is differentiated from the perilymph-containing scala vestibuli and tympani.

In summary, the work of Melhem et al has added to the growing applications of MR imaging of the inner ear. With continued development of higher resolution imaging and sophisticated volumetric analytic techniques, subtle inner ear dysplasia, endolymphatic hydrops, perilymphatic fistula, and progressive cochlear degenerative processes may have reliable imaging correlates. We may soon be able to image and diagnose diseases that to date have eluded radiologic detection.

Patricia A. Hudgins, MD
Member, Editorial Board
Atlanta, GA

References

1. Dahlen RT, Harnsberger HR, Gray SD, et al. **Overlapping thin-section fast spin-echo MR of the large vestibular aqueduct syndrome.** *AJNR Am J Neuroradiol* 1997;18:67–75
2. Schmalbrock P, Dailiana T, Chakeres DW, et al. Submillimeter-resolution MR of the endolymphatic sac in healthy subjects and patients with Meniere disease. *AJNR Am J Neuroradiol* 1996;17: 1707–1716
3. Salt AN, Henson MM, Gewalt SL, Keating AW, DeMott JE, Henson OW Jr. **Detection and quantification of endolymphatic hydrops in the guinea pig cochlea by magnetic resonance microscopy.** *Hear Res* 1995;88:79–86
4. Mark AS, Fitzgerald D. **Segmental enhancement of the cochlea on contrast-enhanced MR: correlation with the frequency of hearing loss and possible sign of perilymphatic fistula and autoimmune labyrinthitis.** *AJNR Am J Neuroradiol* 1993;14:991–996
5. Morris MS, Kil J, Carvlin MJ. **Magnetic resonance imaging of perilymphatic fistula.** *Laryngoscope* 1993;103:729–733

Lymphoma: Master of Chicanery

Lymphoma is a disease with many faces. It seems as if lymphoma is a diagnostic possibility that can, should, or is tagged onto the end of the differential diagnosis in a surprisingly large number of circumstances. Clearly, Chong et al have presented one of these situations in their case report in this issue of the *American Journal of Neuroradiology* (page 1849). An infiltrating mass in the masticator space, with the mandible excluded as a source, in an otherwise healthy patient is far more likely to be a sarcoma (even in an adult), but up pops lymphoma in an appropriate list of differential possibilities.

This case report raises some important practical points in approaching the workup of such mass lesions in the head and neck region, including the following:

1. Whenever an infiltrating mass is present in the extracranial head and neck region, lymphoma is a possible cause, even in the absence of regional adenopathy or other sites of disease.

2. If lymphoma is in the differential diagnosis, so is pseudotumor and sometimes eosinophilic granuloma and Wegener granulomatosis. Even chronic infections with organisms as diverse as blastomycosis and actinomycosis need to be considered when this pattern of disease is present.

3. If lymphoma is the diagnosis, it is almost certainly non-Hodgkin and there is a strong possibility of disease elsewhere in the body requiring an appropriate search for other disease sites.

4. Signal intensity and enhancement patterns present on various pulse sequences are not reliable for distinguishing lymphoma from other malignant neoplasms and from other pathologic processes that may spread in the same anatomic pattern. Tissue sampling in an environment that anticipates lymphoma as a diagnostic possibility is the most efficient path to accurate diagnosis.

5. If the mass is being sampled under imaging guidance and the initial sample shows that lymphoma is likely, several core needle samples should be obtained to save the patient a second needle biopsy or difficult open biopsy, since these masses are often relatively inaccessible by standard surgical approaches.

The inaccessibility mentioned above emphasizes other common sites of origin for this uncommon problem, including the parapharyngeal, retropharyngeal, and buccal spaces as well as the skull base and its immediate environs. The variety in possible sites of origin then predicts that presenting signs and symptoms will be equally diverse. TMJ dysfunction, various cranial neuropathies, dysphagia, otalgia, and a palpable or visible mass are only a few presenting symptoms and signs that, in my experience, eventually proved to be due to lymphoma.

Perhaps one of the more important considerations in the presenting symptoms just listed is that of cranial neuropathy, since the imaging findings can sometimes be subtle. Lymphoma may be "neurotropic," producing only a thickened or enlarged appearance of the lower cranial nerves, which can be traced intracranially when there is continued enlargement of the involved nerves without associated meningeal disease. Once seen, this pattern is hard to forget, and it can lead to a rapid disposition for the patient in what otherwise would prove to be a perplexing and prolonged evaluation.

The more common presentation of lymphoma in the extracranial head and neck is, of course, nodal disease, but even this may be atypical. An isolated mass anterior to the canine fossa of the maxilla is likely to be lymphoma arising in the infraorbital lymph node. This is part of the little-known facial lymph node group that might also be the site of origin or involvement of lymphoma arising over the malar eminence (zygomatic node), in the buccal space (buccal nodes), and along the mandible (mandibular node).

Nodes arising in the more standard parotid and cervical groups must be distinguished from reactive adenopathy and metastases from other malignant lesions. Imaging in these instances may help the clinician to chose the node most accessible for safe, total, excisional biopsy whenever necessary. Whenever nodal disease is present, a search for involvement of extranodal sites is mandatory, and the scanning protocol for the head and neck region must include the orbits (lacrimal glands), Waldeyer's ring, and the entire neck through and including the thyroid gland and nodes in the supraclavicular fossa. This search must include a careful viewing of bone and soft tissues as well as the epidural space and those intracranial structures that happen to be included.

The essence of a good head and neck oncologic imaging examination is a combination of excellent-