History

A 19-month-old boy arrived in the emergency department with a 3-day history of left ear pain and a 1-day history of left facial paralysis. He had been seen by his primary care physician the day before and started on amoxicillin (40 mg/kg/day) for acute otitis media. His facial function was normal at that time. The patient had a history of recurrent otitis media requiring four to five courses of antibiotics per year since birth. Physical examination revealed an alert boy with a temperature of 39°C. Auricular examination was unremarkable. The left external auditory canal was occluded with granulation tissue. Cranial nerve examination demonstrated complete left-sided facial paralysis classified as a House-Brackmann VI paralysis (Table 2.1). The remainder of the head and neck examination revealed no abnormality. A computed tomography (CT) scan was then ordered.

Differential Diagnosis—Key Points

1. Complications of otitis media can be classified according to extracranial and intracranial involvement (Table 2.2). The differential diagnosis of extracranial complications of acute otitis media can be divided into two groups: intratemporal and extratemporal. Intracranial complications include extradural granulation tissue, sigmoid sinus thrombosis, brain or subdural abscess, otitic hydrocephalus, and meningitis. An accurate history, complete otolaryngologic and neurologic examination, and radiographic imaging are needed to differentiate the various complications.

2. All cases of acute otitis media involve inflammation of the mastoid air cells. However, clinically significant acute mastoiditis is a clinical diagnosis based on the findings of suppurative otitis media, postauricular swelling with loss of postauricular crease, and protrusion of the auricle. Coalescent mastoiditis is a specific radiographic diagnosis based on CT and is differentiated from acute mastoiditis by radiographic evidence of loss of the bony septations.

3. Suppurative labyrinthitis occurs when bacterial invasion penetrates the otic capsule, usually via the round window or oval window. The classic presentation is rapid onset of vertigo, sensorineural hearing loss, nausea, and vomiting during an episode of acute otitis media. In the absence of associated meningitis, the cerebrospinal fluid pressure and analysis are normal. Suppurative acute petrositis occurs when there is extension of the middle-ear infection into the petrous
apex, resulting in symptoms of retro-orbital pain, persistent otorrhea, and sixth cranial nerve palsy. This symptom complex is known as the Gradenigo triad. Facial paralysis, usually unilateral, can occur during an episode of acute otitis media either secondary to direct inflammation through a bony dehiscence in the tympanic segment of the facial nerve or secondary to osteitis involving the bony fallopian canal.

4. Extratemporal complications occur when infection progresses to involve the cortical bone surrounding the mastoid air cells. Osteitis of the lateral cortex can result in the development of a subperiosteal abscess. The patient usually presents with more pronounced auricular protrusion, loss of the postauricular crease, and fluctuance over the mastoid. Osteitis of the medial or inferior mastoid cortex can result in the development of a deep neck space infection known as Bezold abscesses.

5. Acute mastoiditis can be accompanied by a significant amount of inflammation of the cartilaginous external auditory canal or

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<tr>
<th>Table 2.1</th>
<th>House-Brackman Facial Nerve Grading System</th>
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<tr>
<td>Grade</td>
<td>Gross</td>
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<td>-----------</td>
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</tr>
<tr>
<td>I. Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>II. Mild dysfunction</td>
<td>Slight weakness on close inspection, slight synkinesis</td>
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<tr>
<td>III. Moderate dysfunction</td>
<td>Obvious but not disfiguring facial asymmetry. Synkinesis is noticeable but not severe. May have hemifacial spasm or contracture</td>
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<tr>
<td>IV. Moderately severe dysfunction</td>
<td>Asymmetry is disfiguring or obvious facial weakness</td>
</tr>
<tr>
<td>V. Severe dysfunction</td>
<td>Only slight, barely noticeable, movement</td>
</tr>
<tr>
<td>VI. Total paralysis</td>
<td>No movement</td>
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<thead>
<tr>
<th>Table 2.2</th>
<th>Complications of Acute Otitis Media</th>
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<tr>
<td>I. Extracranial</td>
<td>A. Intratemporal</td>
</tr>
<tr>
<td></td>
<td>1. Acute mastoiditis</td>
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<tr>
<td></td>
<td>2. Coalescent mastoiditis</td>
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<td></td>
<td>3. Labyrinthitis</td>
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<td>4. Petrositis</td>
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<td>5. Facial nerve dysfunction</td>
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<tr>
<td></td>
<td>6. Tympanic membrane perforation</td>
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<tr>
<td>B. Extratemporal</td>
<td>1. Subperiosteal abscess</td>
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<tr>
<td></td>
<td>2. Bezold abscess</td>
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<tr>
<td>II. Intracranial</td>
<td>A. Extradural granulation tissue and/or abscess</td>
</tr>
<tr>
<td></td>
<td>B. Sigmoid sinus thrombosis</td>
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<td></td>
<td>C. Brain abscess</td>
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<tr>
<td></td>
<td>D. Otic hydrocephalus</td>
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<td></td>
<td>E. Meningitis</td>
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<td>F. Subdural abscess</td>
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granulation tissue, making visualization of the tympanic membrane difficult or impossible in the awake child. This can clinically mimic acute external otitis without middle-ear or mastoid involvement. The physical examination can help differentiate the two entities. Manipulation of the external auditory canal by pulling on the tragus is extremely painful in acute external otitis, but not in mastoiditis. Occasionally, initial response to intravenous (IV) and ototopical treatments is needed to make an accurate diagnosis.

6. Intracranial complications of acute otitis media manifest with a broad range of signs and symptoms. Presentation can range from headache and lethargy to seizures and focal neurologic signs. Sigmoid sinus thrombosis classically presents with “picket-fence” fevers, toxemia, septic embolization, and torticollis. However, further complications can occur with thrombus propagation, including jugular foramen syndrome, otic hydrocephalus, coma, and even death.

7. Brain abscesses can be difficult to diagnose because the signs and symptoms are often subtle. Presentation depends on the stage of the abscess. The first stage, cerebritis, is accompanied by generalized symptoms of headache, malaise, fever, and drowsiness, followed by a quiescent or latent phase that can last for weeks. In the third phase, abscess formation occurs with focal neurologic signs. The final phase, termination, results in rupture of the abscess, leading to rapid deterioration and death.

8. Meningitis presents with classic findings of headache, fever, and neck rigidity. Kernig and Brudzinski signs are positive. Spread of infection to the meninges is by hematogenous dissemination, inner ear malformations (incomplete partitioning), or direct spread (middle fossa dehiscence, meningoencephalocele).

◆ Test Interpretation

The CT scan revealed opacification of the left middle ear and mastoid with normal mastoid trabeculae. There was no bony dehiscence of the tympanic segment of the facial nerve course. The lateral mastoid cortex was intact (Fig. 2.1). CT is the appropriate modality to assess the mastoid and temporal bone. If intracranial involvement is suspected, contrast-enhanced magnetic resonance imaging is a superior investigation. It is more sensitive for the detection of an early brain abscess, sigmoid sinus thrombosis, subdural abscess, and extradural granulation tissue.

◆ Diagnosis

Acute otitis media with facial nerve paralysis

◆ Medical Management

Empirical IV antimicrobial coverage should include coverage for the most common organisms that cause acute otitis media, including Streptococcus pneumoniae, Haemophilus influenzae, and Moraxella catarrhalis. Once culture results are obtained, coverage can be narrowed. Following surgical drainage, symptoms usually dissipate quickly, and oral antibiotic therapy can be instituted. Ciprofloxacin-dexamethasone otic drops should be instilled after placement of tympanostomy tubes. This allows ototopical
treatment of the infection and inflammation. Oral steroid therapy should also be started to address inflammation associated with the facial nerve paralysis.

**Surgical Management**

On the day of presentation, this child underwent bilateral myringotomy and pressure-equalizing tube placement. He was started on IV cefuroxime at the time of surgery in addition to ciprofloxacin-dexamethasone otic drops. Cultures taken at the time of surgery grew penicillin-sensitive S. pneumoniae. By postoperative day 3, facial function had improved, but a significant weakness was still present (House-Brackmann IV). The ear was no longer draining, and the middle ear was dry. The patient was sent home on amoxicillin, otic drops, and a tapering course of oral steroids on postoperative day 4.

Most surgeons believe that a facial nerve paralysis complicating acute otitis media requires urgent surgical drainage of the middle-ear space. The middle ear can be drained using a wide-field myringotomy or pressure-equalizing tube. Whether to address the mastoid is more controversial. When a coalescent mastoiditis is present in a child with extracranial complications of acute otitis media, a cortical mastoidectomy is advised. When mastoid septae are intact, tympanostomy tube placement, medical treatment, and close observation constitute a reasonable initial approach. Any deterioration of the child warrants a mastoidectomy.

**Rehabilitation and Follow-up**

The patient was seen in clinic 1 week later, and facial function had fully recovered. At his last follow-up appointment 6 months after surgery, this child’s pressure-equalizing tubes had extruded, the tympanic membranes were intact, and hearing was normal. He has had one episode of otitis media since this complication.

**Questions**

1. Facial nerve dysfunction characterized by an obvious, but not disfiguring, difference between the two sides, complete eye closure with effort, and normal tone at rest is classified as a House-Brackman:
   A. Grade II
   B. Grade III
   C. Grade IV
   D. Grade V

2. The Gradenigo triad, the symptom complex associated with petrositis, is characterized by all of the following except:
   A. Cranial nerve VII palsy
   B. Retro-orbital pain
   C. Persistent otorrhea
   D. Cranial nerve VI palsy

3. Which of the following is the most common organism causing acute otitis media?
   A. *Haemophilus influenzae*
   B. *Moraxella catarrhalis*
   C. *Streptococcus pneumoniae*
   D. *Staphylococcus aureus*

**Suggested Readings**


A 75-year-old man presents with right-sided hearing loss 2 days before presentation. Although there was no history of trauma preceding the hearing loss, the patient states that he was recovering from a cold. He also describes concomitant, right-sided tinnitus and aural fullness. He denies otorrhea, aural fullness, headache, or vision changes. He denies previous ear infections. He does have a history of left profound sudden hearing loss 10 years ago. His medical history includes hypertension, type 2 diabetes mellitus, coronary artery disease, and hypercholesterolemia. His family history is negative for autoimmune disease or hearing loss.

Neuro-otologic examination is significant for Weber and Rinne tests that the patient could not hear. Microscopic examination of his external auditory canals and tympanic membranes reveals normal anatomy with no evidence of acute infection or sequelae of previous otitis media. His cranial nerves are intact, and there is no spontaneous, gaze-evoked, or positional nystagmus. His cerebellar examination is unremarkable, and his gait and Romberg are normal.

**Differential Diagnosis—Key Points**

Sudden sensorineural hearing loss (SNHL) is defined as a 30-dB or greater sensorineural hearing loss occurring in at least three contiguous frequencies within 3 days or less. The possible causes of sudden SNHL are numerous. Often, the potential causes can be narrowed by the patient’s history and physical examination. It is helpful to consider the possible causes in categories, such as infectious, inflammatory, vascular, neoplastic, traumatic, toxic, and idiopathic.

1. **Infectious.** Many infectious processes can affect the inner ear either unilaterally or bilaterally. Viral infections are commonly implicated, including cytomegalovirus, mumps, influenza, herpes simplex, measles, rubella, and human immunodeficiency virus. An association between upper respiratory tract infections (i.e., viral URIs) and sudden SNHL. Bacterial meningitis or labyrinthitis, syphilis, and mycobacterial or fungal infections may also be implicated. Infections are probably the most common cause of sudden SNHL.
2. **Inflammatory.** Several inflammatory processes can cause sudden SNHL, including autoimmune inner ear disease, Cogan syndrome, Wegener granulomatosis, polyarteritis nodosa, rheumatoid arthritis, systemic lupus erythematosus, and other rheumatologic disorders. These immune-mediated etiologies can be isolated to the inner ear (primary) or may be due to systemic autoimmune disease (secondary).

3. **Vascular.** Occlusion of the labyrinthine artery may result in sudden SNHL. Other vascular diseases—including leukemia, sickle cell disease, vasculitis, and embolization during coronary bypass surgery—can cause acute SNHL.

4. **Neoplastic.** Multiple tumors may directly or indirectly affect the inner ear and result in SNHL. About 13% of patients with acoustic neuromas present with sudden SNHL. Other tumors that can cause SNHL include meningioma and other cerebellopontine angle tumors, facial neuromas, squamous cell carcinoma of the external auditory canal or middle ear, glomus tumors, and papillary adenocarcinoma of the endolymphatic sac.

5. **Traumatic.** Penetrating or blunt trauma to the temporal bone can result in SNHL through direct damage to the labyrinth or internal auditory canal, subluxation of the stapes, or concussive injury.

6. **Toxics.** Several ototoxic medications can result in SNHL. The most commonly encountered toxins include aminoglycosides, loop diuretics, and cisplatin. These agents commonly cause bilateral SNHL, but unilateral loss may occur.

7. **Miscellaneous.** Other possible causes of sudden SNHL include diabetes mellitus, perilymphatic fistula, cochlear hydrops, congenital inner-ear deformities, and operative complications of otologic surgery.

**◆ Test Interpretation**

Investigation into the cause of this hearing loss should include serologic testing with complete blood count, erythrocyte sedimentation rate, C-reactive protein, coagulation studies, thyroid function studies, lipid profile, and fluorescent treponemal antibody absorption. Additional testing can include rheumatoid factor, antinuclear antibody, heat-shock protein 70 antibody, and Lyme titers.

Audiometric testing, including pure-tone thresholds, speech reception and discrimination, and acoustic reflexes, is mandatory. In this case, the audiogram reveals profound bilateral sensorineural loss (Fig. 8.1). Magnetic resonance imaging with the internal auditory canal–cerebellopontine angle with contrast is required to rule out a retrocochlear lesion, such as an acoustic neuroma.

**◆ Diagnosis**

Unilateral right-sided SNHL in a patient with a history of prior contralateral profound, sudden SNHL.

**◆ Medical Management**

Although the natural history is variable, many patients with sudden SNHL experience spontaneous improvement within the first 2 weeks. The treatment regimens are numerous. However, the most widely accepted treatment is corticosteroids, administered systemically (orally or intravenously) or transtympanically. If systemic steroid therapy is undertaken, close attention must be paid to systemic side effects, such as elevated blood sugars (worsening diabetes management), elevated blood pressure, and aseptic femoral head necrosis; prophylaxis for gastric ulcers must be administered.

Additional treatment modalities have been used less commonly, including vasodilators, inhaled carbogen, anticoagulants, and antivirals.

**◆ Surgical Management**

Surgical management may be indicated in the setting of a retrocochlear mass lesion. Cochlear implantation may be offered in the setting of bilateral profound hearing loss.

**◆ Rehabilitation and Follow-up**

This patient will require close monitoring of blood glucose levels and medical status during the period of oral steroid use. Follow-up
audiometry as well as testing to exclude retrocochlear pathology must be obtained. If this patient has no return of hearing, cochlear implantation would be appropriate.

Questions

1. What is the most common cause of sudden sensorineural hearing loss?
2. What study is required of all patients with sudden sensorineural hearing loss to rule out a retrocochlear process?
3. By what two routes have steroids been administered for sudden SNHL?

Suggested Readings


Multinodular Goiter

David L. Steward

**History**

A 56-year-old woman with a longstanding palpable multinodular goiter is referred by her primary care physician for evaluation. The patient reports a family history of goiter and thyroid surgery but no family history of thyroid cancer. She denies history of radiation exposure. She denies dysphagia, dyspnea, or hoarseness. She denies symptoms of hypothyroidism or hyperthyroidism.

Physical examination reveals multinodular goiter with around 3-cm right-sided and 2-cm left-sided dominant nodules. Vocal cord function is normal. No other masses or evidence of adenopathy are noted.

**Test Interpretation**

Ultrasound is the best imaging modality for thyroid nodules to assess the presence of non-palpable nodules, for accurate sizing, to identify features suspicious for malignancy, and to guide fine-needle biopsy. Suspicious sono-graphic features of thyroid nodules have an 80% sensitivity for malignancy. Size alone is nondiscriminatory, but larger nodules (>1.5–2 cm) are often biopsied to exclude malignancy even in the absence of suspicious features. In the presence of multiple nodules, biopsy of suspicious nodules should be preferentially performed over benign-appearing larger ones. Often the largest on each side is biopsied along with any sonographically suspicious nodules, requiring biopsy of as many as four nodules to exclude malignancy within a large multinodular gland (see papillary thyroid carcinoma case for further description of ultrasound-guided fine-needle biopsy and cytologic findings).

Screening thyroid-stimulating hormone (TSH) should be done to exclude subclinical hyperthyroidism, which is common in multinodular goiter. If the TSH is low, then follow-up testing of free tri-iodothyronine (T₃) and free thyroxine (T₄) is necessary to confirm hyperthyroidism from toxic multinodular goiter.

**Differential Diagnosis—Key Points**

Multinodular goiter is common and often hereditary. Three main points come up when evaluating a patient with multinodular goiter: (1) Does this represent malignancy? (2) Is there endocrine dysfunction? (3) Is there evidence of compression of surrounding structures as a result of its size?
Slowly growing multinodular goiters confined to the neck are often asymptomatic to the patient as the neck musculature and skin expand with the growth. Goiters that extend into the thoracic inlet and mediastinum more commonly cause tracheal deviation and compression requiring chest computed tomography for further evaluation. Pulmonary function tests in the sitting and supine position or with flexion and extension may be helpful to assess dyspnea. Barium swallow to evaluate dysphagia rarely demonstrates extrinsic compression but may be helpful to exclude other causes of dysphagia, such as esophageal dysmotility or strictures, especially in older individuals.

The TSH level is normal, suggesting a euthyroid state. Ultrasound confirms a multinodular goiter with solid dominant hypoechoic nodules bilaterally, 2.1 × 2.4 × 3.2 cm on the right and 1.8 × 1.5 × 2.5 cm on the left side. Numerous nonpalpable subcentimeter nodules are noted, along with a left superior pole hypoechoic nodule measuring 1.4 × 1.5 × 2.1 cm and partially cystic right inferior hypoechoic nodule measuring 1.3 × 1.4 × 2.1 cm. No evidence of nodal disease or extrathyroidal extension is seen. None of the nodules contains multiple suspicious features for malignancy.

**Diagnosis**
Multinodular goiter

**Medical Management**

Medical management generally involves fine-needle biopsy to exclude malignancy and observation for significant interval growth. This patient underwent fine-needle biopsy of all four nodules without evidence of malignancy and elected for observation.

Thyroid hormone replacement therapy to keep TSH levels in the low normal range between 1 and 3 is indicated for patients with hypothyroidism. Thyroid hormone suppression therapy with levothyroxine to suppress TSH to subnormal values may reduce goiter size for some patients but is associated with increased risk of cardiac arrhythmia and osteoporosis, and goiter recurs following discontinuation of suppressive therapy. Routine suppression is not recommended.

Radioiodine may be more effective than TSH suppression, but it requires higher doses of radioiodine for euthyroid multinodular goiter compared with patients with hyperthyroidism. Radioiodine may reduce volume size by 20 to 40% over a 1- to 2-year period but is associated with a small risk of malignancy and is not recommended except possibly for older or medically infirm patients with compressive symptoms who do not want to undergo surgery. TSH-stimulated radioiodine therapy may improve radioiodine uptake and subsequent volume reduction with lower radioiodine doses and should be considered if this mode of therapy is undertaken.

**Surgical Management**

Total thyroidectomy is curative for multinodular goiter that is bilateral. In unilateral nodular goiter, fine-needle biopsy to exclude malignancy followed by hemithyroidectomy will be sufficient for most patients but may require completion thyroidectomy if occult malignancy is discovered or subsequent enlarging nodular disease develops in the remaining lobe. Partial lobectomy, subtotal thyroidectomy, and bilateral subtotal thyroidectomy should be avoided because of the likelihood of recurrent nodular disease requiring reoperative surgery in a prior operative field with increased risk. Surgery should be reserved for cases suspicious for malignancy or for large or enlarging goiters, especially in younger patients. Involvement of the thoracic inlet or superior mediastinum or evidence of tracheal compression or significant deviation is indication for surgery.

**Rehabilitation and Follow-up**

Annual TSH testing and annual to biannual ultrasound are reasonable to evaluate for progression of nodular disease, development of new nodules, or development of thyroid dysfunction. This patient has demonstrated no significant disease progression or symptoms.
in long-term follow-up. Repeat fine-needle biopsy to exclude malignancy in previously cytologically benign nodules without significant change on ultrasound is not necessary but should be considered if suspicious sonographic features appear or significant growth occurs.

◆ Questions

1. What is the best imaging modality for a euthyroid patient suspected of having a thyroid nodule?
   A. Ultrasound
   B. CT scan
   C. MRI scan
   D. Radioiodine uptake and scan
   E. PET scan

2. What biochemical screening test is indicated for a patient suspected of having a multinodular goiter?
   A. Thyroglobulin
   B. Calcitonin
   C. Thyroid-stimulating hormone
   D. Free thyroxine
   E. Free thryonine

3. What is the best surgical procedure for management of an enlarging bilateral multinodular goiter with compressive symptoms?
   A. Bilateral subtotal thyroidectomy
   B. Unilateral subtotal thyroidectomy
   C. Total thyroidectomy
   D. Hemithyroidectomy (lobectomy)
   E. Isthmusectomy

Suggested Reading

Cooper DS, Doherty GM, Haugen BR, et al, for the American Thyroid Association Guidelines Taskforce. Management guidelines for patients with thyroid nodules and differentiated thyroid cancer. Thyroid 2006;16(2):1–33