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Mitchell S.V. Elkind,
MD, MS

Clinical Reasoning: A 50-year-old woman with deep stabbing ear pain

Justin M. DeLange, DO
Ivan Garza, MD
Carrie E. Robertson, MD

Correspondence to
Dr. Robertson:
robertson.carrie@mayo.edu

SECTION 1

A 50-year-old woman presented with deep ear pain on the right that started 3 days prior. She described the pain as aching with superimposed severe stabbing. This pain was associated with hyperacusis, but she otherwise denied any hearing loss, muffled hearing, tinnitus, diplopia, dysphagia, facial weakness, numbness, or rash.

Questions for consideration:

1. What would be the differential diagnosis of this patient's acute ear pain at this point in her care?
2. Which nerves carry sensation from the ear (and can therefore refer pain to the ear)?

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From the Department of Neurology, Mayo Clinic, Rochester, MN.

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Figure Sensory innervation of the ear and surrounding structures

Depiction of the sensory nerves shows the innervation of the ear and surrounding anatomy. Each box with its corresponding color illustrates each nerve's distribution. Sensory distributions may overlap. Illustration by John Hagen, Mayo medical illustrator. Used with permission of Mayo Foundation for Medical Education and Research; all rights reserved.

Table Secondary (referred) causes of otalgia				
CN VII (nervus intermedius)	CN V (auriculotemporal nerve)	CN IX (Jacobson nerve)	CN X (Arnold nerve)	C2, C3 (great auricular nerve and lesser occipital nerve)
Cerebellopontine angle tumors	Temporomandibular dysfunction (TMJ disease)	Pharyngitis	Laryngopharyngeal reflux (GERD)	Cervical degenerative disease/arthritis
Herpes zoster	Dental pathology	Tonsillitis/peritonsillar abscess	Laryngeal tumor/cancer	Whiplash/trauma
Nervus intermedius neuralgia	Parotiditis	Post-tonsillectomy	Thyroid tumor/inflammation	Cervical meningiomas
	Parotid tumor	Pharyngeal tumor/cancer (SCC)	Intrathoracic mass lesion	Cervical lymphadenitis
	Oral cavity cancer (SCC)	Retropharyngeal/parapharyngeal abscess	Laryngitis	Great auricular/lesser occipital neuralgia
	Acute sinusitis	Acute sinusitis (if direct invasion of middle ear)	Arnold neuralgia	
	Trigeminal neuralgia	Eagle syndrome; laryngopharyngeal reflux (GERD)		
		Glossopharyngeal neuralgia		

Abbreviations: CN = cranial nerve; GERD = gastroesophageal reflux disease; SCC = squamous cell carcinoma; TMJ = temporomandibular joint.

SECTION 2

The differential for otalgia can be broadly divided into primary otalgia (related to pathology within the ear) and secondary (referred) otalgia. Primary etiologies involve pathology anywhere along the external, middle, and inner ear, and include the following:

- Infectious and inflammatory etiologies: otitis media, otitis externa, herpes zoster oticus (Ramsay-Hunt syndrome), mastoiditis, myringitis (inflammation of the tympanic membrane), and cellulitis/chondritis
- Trauma: blunt/penetrating trauma, foreign body, barotrauma, tympanic membrane perforation, and auricular hematoma
- Neoplasm: cholesteatoma, adenocarcinoma, squamous cell carcinoma

A patient with new otalgia should receive a detailed examination with otolaryngology. If unremarkable, then nonotologic or referred sources of ear pain should

be considered. Nerves referring pain to the ear include branches of cranial nerves V, VII, IX, and X, and upper cervical roots (figure). The table lists some specific causes of referred pain to the ear.¹

Four days after the onset of her ear pain, the patient developed subacute right-sided facial weakness with difficulty raising her eyebrow, closing her eye, smiling, and keeping her mouth closed on the right. She also noted that food tasted different on the right. Her extraocular movements, including abduction in each eye, were full. Formal evaluation with otolaryngology showed no evidence of ear pathology, including no evidence of lesions or vesicles in the external auditory canal.

Questions for consideration:

1. What is the differential diagnosis for the new facial weakness?
2. What investigations and treatment would you consider at this point?

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SECTION 3

The most common cause of isolated facial neuropathy is Bell palsy, with an incidence of about 20 per 100,000 annually.² Symptoms tend to come on suddenly (over hours to a day), reaching maximal facial weakness within 5 days of the onset.³ Many, but not all, patients report a prodromal upper respiratory illness.³ Bell palsy is considered idiopathic, but there seems to be an association with herpes simplex virus. Ramsay-Hunt syndrome can present similarly and is caused by reactivation of herpes zoster in the geniculate ganglion of the facial nerve. Ramsay-Hunt syndrome tends to have more severe facial weakness, vesicular eruption around ear and throat, and may involve the eighth cranial nerve, with associated deafness and vestibular dysfunction. Acute isolated facial neuropathy can also be caused by cytomegalovirus, Epstein-Barr virus, adenovirus, rubella virus, mumps, influenza B, coxsackievirus, Lyme disease, HIV, and neurosarcoidosis.⁴ Gradually progressive facial neuropathies can be caused by masses anywhere along the course of the peripheral facial nerve, including the cerebellopontine angle, facial canal, and parotid gland.

The evaluation for Bell palsy includes a detailed neurologic examination, including a bedside test of hearing and full extraocular movements. Patients commonly have accompanying hyperacusis and dysgeusia ipsilaterally. The ear canal, tongue, and pharynx should be examined for herpes zoster vesicles, though these may not show for up to 2 weeks after the palsy.⁵

Red flags for nonviral etiologies include slowly progressive symptoms, a lack of improvement after 6 months, bilateral facial paresis, significant involvement of other cranial nerves, and associated systemic illness.^{4,6} If these are present, consider a contrast-enhanced brain MRI, with special attention to the

facial nerve including the parotid gland. A CT of the temporal bone can help assess the structure of the facial canal. An electromyogram with blink reflexes can help assess the degree of facial neuropathy and identify trigeminal nerve involvement. Serologic testing for Lyme, antinuclear antibody, SSA/SSB, erythrocyte sedimentation rate, angiotensin converting enzyme, and hemoglobin A1C could be considered. A chest X-ray for hilar lymphadenopathy can be performed if neurosarcoidosis is suspected. If more than one cranial nerve is involved, or if the course is progressive without clear etiology, a lumbar puncture for glucose, protein, cells/cytology, Lyme serology, West Nile virus serology, oligoclonal bands, and viral cultures could be performed.

Treatment guidelines for Bell palsy recommend a course of oral glucocorticoids as there is good evidence to suggest facial recovery. The addition of antiviral agents such as acyclovir or valacyclovir is often considered although their benefit has not been established. Antiviral agents typically have a modest effect in recovery of facial function.²

The patient underwent steroid therapy for her facial palsy with good motor function recovery. However, 2 years later, she still had severe intermittent right ear pain, described as a deep ache with superimposed stabbing pains deep within the ear, sometimes with radiation to the right mandibular angle. She continued to deny any rashes or vesicular lesions on her face, head, neck, or ears. On examination, she had mild residual right facial weakness with subtle flattening of nasal labial fold. Stabbing pain could be elicited by pressure on the right anterior ear canal with a cotton swab. Neurologic examination was otherwise unremarkable.

Questions for consideration:

1. What would the diagnosis be now?
2. What treatment can be offered for the pain?

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SECTION 4

At this point, the diagnosis is most consistent with nervus intermedius neuralgia (NIN), also called “geniculate neuralgia.” The nervus intermedius is a small branch of the facial nerve that carries sensory information from the ear, tympanic membrane, and acoustic meatus. It also carries parasympathetic information to the lacrimal and salivary glands and taste sensation from the anterior two-thirds of the tongue.⁷

NIN is characterized by severe paroxysmal neuralgic pain in the auditory canal. The International Headache Society has recommended the following diagnostic criteria: recurring pain in paroxysmal attacks lasting from a few seconds to minutes; pain is severe in intensity and shooting, stabbing, or sharp in quality; and the presence of a trigger zone in the posterior wall of the auditory canal or periauricular region. Disorders of lacrimation, salivation, and taste may accompany the pain.⁸

NIN can be idiopathic or secondary to an underlying cause. Symptomatic (secondary) NIN can be due to many of the same etiologies that are associated with acute facial neuropathy, including Ramsay-Hunt syndrome, Bell palsy, and Lyme disease.⁹ Some previously labeled idiopathic cases of NIN have been noted to have microvascular compression of the nervus intermedius. These select cases may respond to microvascular decompression (mobilization) of the anterior inferior cerebellar artery.⁷

For pain management, carbamazepine should be tried first at the dosage utilized for trigeminal neuralgia. Other agents including oxcarbazepine, lamotrigine, phenytoin, and baclofen can be tried if carbamazepine is ineffective.¹⁰ In idiopathic NIN, neurosurgery is often considered when pharmacotherapy fails. Multiple procedures have evolved over time in hopes of treating this painful disorder. These have included geniculate ganglion resection; nervus intermedius resection; exploration or sectioning of cranial nerves V, IX, and X; and microvascular decompression of cranial nerves V, IX, and X. However, no prospective data exist on the efficacy of these surgical modalities. Thus surgery should be utilized as a last resort.⁷

DISCUSSION When a patient presents with otalgia and normal ear examination findings, localization and diagnosis are often difficult, due in part to the overlapping sensory innervation from multiple nerves. Our patient went on to develop facial nerve palsy, validating the localization to the sensory portion of the facial nerve, the nervus intermedius. In patients with otalgia who do not have such helpful clues on the neurologic examination, we recommend considering the mnemonic ENT: E = ear pathology (intrinsic); N = neuralgia of one of the nerves to the ear (cranial nerves V, VII, IX, X, great auricular nerve,

and lesser occipital nerve); T = structures starting with T (teeth, temporomandibular joint [TMJ], throat, tonsils, tongue, thyroid, trachea, tunnels for vessels [jugular foramen and carotid sheath]).

After a thorough evaluation by Otolaryngology for any otogenic causes of pain, we recommend considering evaluation for pathology related to surrounding structures as listed above, as well as the lateral neck, upper cervical roots, and parotid gland. Brain MRI will not include many of the areas that can refer pain to the ear. In some cases, MRI of the face (including TMJ, parotid, and pharynx) and soft tissues of the neck (to assess thyroid, carotid sheath, and jugular foramen) can be considered. MRI of the internal auditory canal and magnetic resonance angiography of the head with contrast may also be useful in specific cases. If these are negative and the pain is sharp and paroxysmal, consider neuralgia of one of the innervating nerves. Triggers can be helpful in localizing which nerve may be involved. For instance, glossopharyngeal neuralgia is classically triggered by swallowing, auriculotemporal neuralgia may be associated with movement of the TMJ, and nervus intermedius neuralgia is classically triggered by touching the posterior auditory canal.

AUTHOR CONTRIBUTIONS

Dr. DeLange was responsible for drafting/revising the manuscript for content, including medical writing for content, study concept/design, and analysis/interpretation of data. Dr. Garza was responsible for drafting/revising the manuscript for content, including medical writing for content. Dr. Robertson was responsible for drafting/revising the manuscript for content, including medical writing for content, study concept/design, and analysis/interpretation of data.

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