Facial movement and expression disorders

Part 1. Facial weakness

The extraordinary expressive range of the human face is taken for granted in daily life. We invest our faces and those of our loved ones with powerful emotional overtones, and diseases which produce facial disfigurement are uniquely distressing to patients. In Part 1 of this article, key features and underlying causes of facial weakness are outlined.

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Clinical disorders of facial movement may occur at any level of the motor hierarchy: cortex, basal ganglia and other subcortical structures, upper or lower motor neuron, neuromuscular junction and muscle. They can be classified according to the type of motor activity they produce.

In this two-part article, we present frameworks for the diagnosis and management of three different patterns of clinical features:

- unilateral facial weakness (Part 1)
- bilateral facial weakness (Part 1)
- excessive facial movement (Part 2, next month).

An overview of facial neuroanatomy is useful for classifying and understanding the disorders, and is presented in the box on page 53.

Unilateral facial weakness

The diagnosis and management of unilateral facial weakness are outlined in Table 1.

Associated features

Abnormal brainstem signs. If the facial nucleus or intrapontine facial fibres are damaged, it is likely that other evidence of a brainstem lesion will be present, such as an ipsilateral gaze palsy, nystagmus, facial sensory loss or crossed long tract signs.

Deafness. Lesions in the posterior fossa, cerebellopontine angle or petrous temporal bone will tend to produce ipsilateral deafness because of the close proximity of the vestibulocochlear nerve.

Reduced corneal reflex. The corneal reflex may be lost and ipsilateral cerebellar signs may be present with lesions of the cerebellopontine angle. The corneal reflex can be assessed when ipsilateral paralysis of eyelid closure is present by asking the patient if a stimulus is felt, and by observing the normal consensual closure of the opposite eyelid and the upward roll of the eyeball when the cornea is touched on the para-

IN SUMMARY

- **Bell's palsy should not be diagnosed if deafness, a reduced corneal reflex or ptosis is present. However, altered taste or hyperacusis is an important pointer to this diagnosis.**
- **A viral prodrome is present in 60% of cases of Bell's palsy. The strongest association is with herpes simplex.**
- **Unusual or sinister causes should be suspected in all cases of bilateral facial weakness.**
- **Pathological affect should be distinguished from other situations in which emotional lability and euphoria result from diffuse brain pathology or pathological emotions of psychosis or drug intoxications.**

lysed side. The last sign ('Bell's phenomenon') is also apparent when the patient attempts to close the eye forcefully and is a normal event, albeit usually invisible.

Ptosis. Ptosis is not found with isolated facial nerve lesions and, if present, may indicate a Horner's syndrome or an oculomotor nerve lesion.

Altered taste and hyperacusis. Bell's palsy should not be diagnosed if deafness, reduced corneal reflex or ptosis is present. On the other hand, altered taste or hyperacusis is an important pointer to this diagnosis.

Clinical patterns

Bell's palsy is the most common cause of unilateral facial weakness; however, 'all that palsies is not Bell's'.

Bell's palsy

The incidence of Bell's palsy has been estimated at 15 to 40 cases per 100,000, and is not influenced by age or race. The risk is increased fivefold in diabetes and threefold in pregnancy.

Although Bell's palsy is traditionally described as idiopathic, the mechanism is probably acute viral inflammatory demyelination of the facial nerve, which would account for the swelling of the nerve within the facial canal and secondary ischaemia found in some cases. It remains a diagnosis of exclusion, and patients should be followed until the condition resolves or fresh clues to a specific cause appear. All cases of suspected Bell's palsy should be followed to ensure that resolution occurs and new features do not develop.

Presenting features

Bell's palsy is often heralded by pain behind the ear, which usually evolves over 48 hours before reaching a plateau, succeeded by unilateral facial weakness (Figure 1). Although many patients complain of facial numbness, objective sensory testing should be normal.

Taste may be altered and there may be increased sensitivity to sound or hyperacusis (such as when using the telephone). In addition to unilateral facial weakness, excessive tearing may be noted, caused by weakness of the orbicularis oculi (which normally holds the lacrimal puncta against the conjunctiva).

Viral associations

A viral prodrome is present in 60% of cases. The strongest association is with herpes simplex but Bell's palsy has also been reported after:

- chickenpox
- mumps
- Epstein–Barr virus
- cytomegalovirus
- coxsackievirus
- influenza
- HIV.

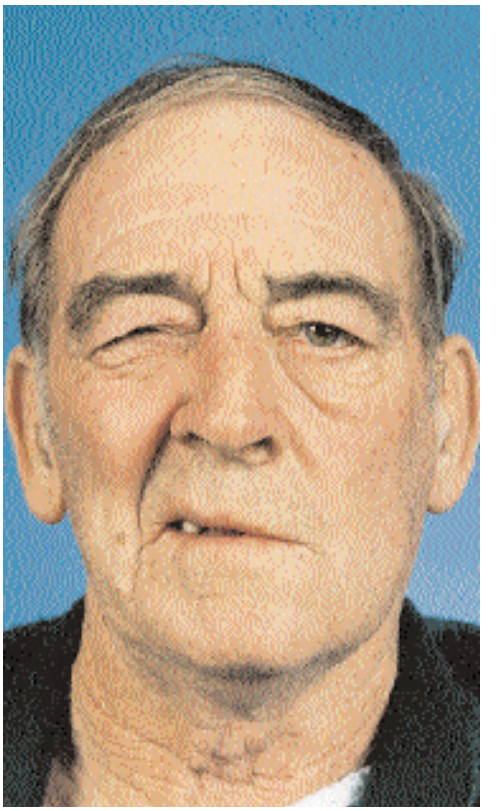

Figure 1. A 69-year-old man with an incomplete left Bell's palsy. Note equal involvement of the upper and lower face, affecting brow wrinkling and eye closure as well as smiling. Facial weakness in lower motor neuron disorders tends to be more severe than in upper motor neuron lesions.

Treatment

Corticosteroids. The use of corticosteroids in Bell's palsy is a controversial area in neurology, which in part reflects the good prognosis of the untreated condition. Despite extensive study, a beneficial effect of steroid therapy in influencing long term outcome has not been proved.

However, a number of studies have suggested that corticosteroids may:

- prevent denervation in cases of complete paralysis
- prevent progression of incomplete to complete paralysis
- shorten the time to recovery
- retard development of abnormal synkinesias.

Corticosteroids are very effective in relieving the pain that is sometimes an early feature of Bell's palsy. It is likely that if benefit is to occur, steroids must be used early in the course (within a week of onset). Although dosage schedules vary, 25 mg of oral prednisolone (Panafcortelone, Solone) daily for a week is one useful regimen, with patient review on completion.

Corticosteroids should not be used when contraindications exist, such as diabetes, hypertension, peptic ulcer disease, osteoporosis, glaucoma or pregnancy.

Other treatment. There is no proven place for adjunctive therapies or surgical decompression of the facial nerve in Bell's palsy. The role of

Table 1. Unilateral facial weakness: diagnosis and management

Signs and symptoms	Site of lesion	Possible diagnoses	Investigations	Management
Weakness of the upper and lower face				
Gaze palsy, hemiparesis, facial numbness	Brainstem	Stroke, pontine glioma	Brainstem MRI	Referral
Reduced corneal reflex, deafness, ataxia	Cerebellopontine angle	Acoustic neuroma, meningioma	Brain MRI	Referral
Vesicles (external ear canal and soft palate), deafness	Geniculate ganglion	Herpes zoster (Ramsay Hunt syndrome)	Herpes serology	Eye care, observation, antiviral therapy (aciclovir [Acyclo-V, Zovirax Tablets, Zyclir Tablets], famciclovir [Famvir] or valaciclovir [Valtrex])
Altered taste, hyperacusis	Facial canal	Bell's palsy	–	Eye care, observation, corticosteroids (if no contraindications)
Otitis, trauma, steadily worsening weakness	Petrous temporal	Infection, trauma, tumour	Cranial CT, temporal bone MRI	Referral
Weakness of the lower face				
Spared emotional smile	Cortex or corticofacial fibres	Stroke, tumour, trauma	Cranial CT, brain MRI	Referral
Localised facial weakness				
Parotid mass, trauma	Facial nerve branches	Parotid tumour, trauma, leprosy	–	Referral

electrical testing to select patients for surgery is now limited, since surgical decompression of the facial nerve has fallen into disfavour, and is not necessary in incomplete palsy.

Complications

The most feared complication of Bell's palsy is exposure keratitis, which occurs if the cornea is not adequately protected. It can be avoided by using artificial tears, instilling lubricating paraffin ointment (Duratears, Lacri-Lube, Poly Visc) and taping (rather than padding) the eye closed at night. Dark glasses should be worn outdoors.

Ophthalmological advice should be sought if the patient reports eye discomfort or the eye becomes irritated despite the above measures. Tarsorrhaphy is rarely necessary in co-operative patients, use of botulinum toxin type A (Botox) to weaken the eyelid levator may be considered if conservative measures fail.

Prognosis

The most favourable prognostic sign is an incomplete rather than complete facial palsy. If the weakness is severe or complete, recovery commencing within three weeks is a favourable sign.

Between 60 and 80% of patients make a complete recovery, particularly if the palsy is incomplete. In these cases, recovery usually begins within eight weeks and is complete by six to 12 months.

How are facial movements and expression controlled?

The corticofacial pathways

The facial motor nucleus, the final common destination of the multiple pathways from the higher centres that mediate human facial expression, is located in the lower pons (Figure A). In general, the lower part of the facial nucleus (which controls the lower face muscles), is supplied by the contralateral cortex; the upper part of the nucleus, (which controls the upper face), is supplied from both cerebral hemispheres.

It follows that a single lesion in the corticofacial pathway from cortex to facial nucleus produces an asymmetrical smile, but relatively preserved forehead wrinkling and eye closure. In contrast, the muscles of the upper and lower face are weakened equally by a lesion of the facial nucleus itself or the facial nerve after leaving the nucleus. This is the basis for the fundamental bedside distinction between upper and lower motor neuron patterns of facial weakness: an upper motor neuron lesion affects the contralateral lower face, a lower motor neuron lesion affects the entire face ipsilateral to the lesion.

Some exceptions to the rule exist. In practice, an acute upper motor neuron lesion often produces some upper facial weakness and a recovering facial nerve lesion may appear to affect smiling more than eye closure. A local lesion of the facial nerve (such as trauma or a tumour) after it divides on entering the face also spares the upper facial muscles. Detection of a mild facial palsy is sometimes difficult because minor asymmetries are common in healthy people. However, a diminished spontaneous blink on the affected side, inability to bury the eyelashes or loss of the ability to wink or whistle are sensitive signs of true facial weakness.

The course of the facial nerve

After leaving the brainstem, the facial nerve pursues a long and tortuous course, through the petrous temporal bone, making it vulnerable to injury at a variety of sites. It supplies all the muscles of facial expression (except the elevators of the eyelids), and conveys:

- taste fibres to the anterior two-thirds of the tongue (via the chorda tympani)
- secretory fibres to the salivary and lacrimal glands
- a small number of sensory fibres from the external auditory canal and retroauricular region.

The facial nerve also supplies the small stapedius muscle that damps excessive oscillations of the tympanic membrane in response to loud sounds.

These other functions of the facial nerve are often spared in nerve lesions but they permit more accurate localisation of the site of damage if affected. As a guide, if altered taste, lacrimation or hearing (hyperacusis) is present, the lesion must lie within or proximal to the petrous temporal bone.

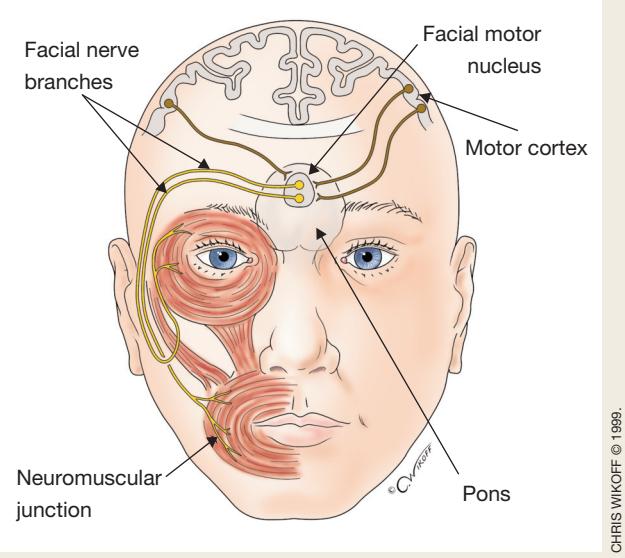

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Figure A. Facial motor pathways. Voluntary and posed facial expressions are controlled by the cerebral cortex, mediated by subcortical structures that project into the facial motor nucleus. The final common motor pathway is the facial nerve. Impaired or excessive facial movement may result from lesions at any level of this hierarchy.

The muscles of facial expression

The 17 paired muscles of facial expression, innervated by the facial nerve, differ from other skeletal muscles in several important respects. They have a distinct origin in embryonic life and retain some autonomic connections via the facial nerve, emphasising their potential to respond to emotional states. They develop within the moveable facial skin and fascia, without bony attachments and participate in specialised cutaneous reflexes, including the protective blink reflex, which can be triggered by stimuli delivered to a wide area of the face.

Early theories of facial expression assigned a characteristic emotion to each muscle. This is overly simplistic, but the action of particular muscles can dramatically alter the emotional message conveyed, a point that is well understood by artists and portrait photographers. The most famous example is the contraction of the orbicularis oculi in emotional (but not posed) smiling. As Duchenne put it in 1862, this muscle 'is only put in play by the sweet emotions of the soul'.

Table 2. Bilateral facial weakness: diagnosis and management

Signs and symptoms	Site of lesion	Possible diagnoses	Investigations	Management
Impaired voluntary movements with ptosis				
Involvement of other muscles	Muscle	Myotonic and other dystrophies, polymyositis	Electromyography, muscle biopsy	Referral
Fatiguability	Neuromuscular junction	Myasthenia gravis	Tensilon test, electromyography	Referral
Impaired voluntary movements without ptosis				
Involvement of other cranial nerves	Cranial polyneuropathy	Cancer, sarcoid, HIV, TB	Skullbase MRI, CSF examination	Referral
Generalised weakness, hyporeflexia	Peripheral nerve	Guillain–Barré syndrome	Nerve conduction studies, CSF examination	Referral
Pseudobulbar palsy, pathological affect	Corticofacial fibres	Multiple sclerosis, motor neuron disease, stroke	Brain MRI	Trial of tricyclic antidepressant, referral
Impaired emotional expression				
Limb tremor, rigidity, bradykinesia, falls, gaze palsy	Basal ganglia	Parkinson's disease and related syndromes	–	Trial of levodopa, referral

The remainder of patients show varying degrees of residual effects, including:

- facial weakness
- 'jaw winking' and other abnormal associated facial movements (synkinesias)
- 'crocodile tears' (rare) – an excessive flow of tears when eating, caused by the aberrant reinnervation of the lacrimal gland by regenerating facial nerve fibres.

The longer the delay in return of movement, the poorer the recovery will be. Other adverse prognostic features include:

- older age
- hyperacusis
- decreased tearing
- pain in the ear canal or face
- associated hypertension, diabetes or psychiatric illness.

Approximately 10% of patients will have recurrent facial palsy. In this situation, alternative causes should be excluded (such as diabetes, tumours, sarcoidosis or infection). A family history is present in approximately 10% of patients.

Ramsay Hunt syndrome

Herpes zoster facial paresis may be associated with severe otalgia, hearing loss and vesicles of the external ear canal and soft palate, which should be sought in all cases of apparently idiopathic facial palsy. This has been designated Ramsay Hunt syndrome, and carries a worse prognosis.

An oral antiviral agent (aciclovir [Acyclo-V, Zovirax Tablets, Zyclir Tablets], famciclovir [Famvir], valaciclovir [Valtrex]) should be given to these patients.

Other causes of unilateral facial weakness

Atypical presenting features or a progressive course will require further investigation to exclude sinister causes of facial weakness, most of which will require specialist referral. These are discussed in Table 1.

Bilateral facial weakness

Underlying causes

Bilateral facial weakness (often asymmetrical) should prompt a search for evidence of unusual disorders.

On careful physical examination, there is usually evidence elsewhere of the underlying disorder, such as:

- Guillain–Barré syndrome
- carcinoma (see Figure 2)
- infectious mononucleosis
- HIV
- tuberculous meningitis
- sarcoidosis
- other causes of mononeuritis multiplex
- myasthenia gravis (see Figure 3)
- myopathy.

The diagnosis and management of bilateral facial weakness are discussed in Table 2.

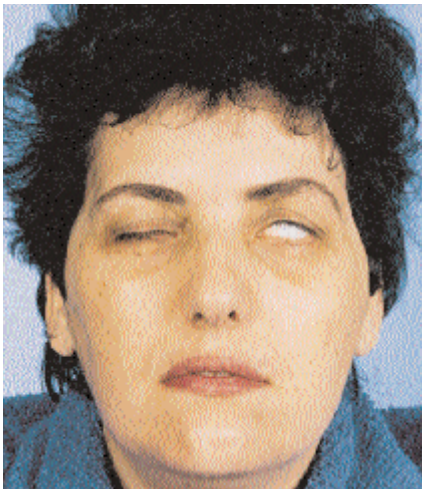


Figure 2. A 41-year-old woman with bilateral facial weakness caused by carcinomatous meningitis with an unknown primary. Both facial nerves are involved, producing a lower motor neuron pattern of weakness on both sides. Note the failure to bury the eyelashes on the less severely affected right side. Upward roll of the eyeball on attempted lid closure, which is normally invisible, is clearly seen on the left eye.

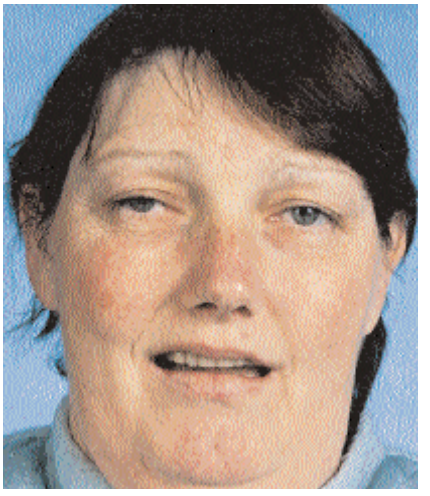


Figure 3. A 40-year-old woman with myasthenia gravis. Bilateral ptosis is evident (more severe on the right) and a weak horizontal smile. Ptosis is an important clue to a myopathic process, and in myasthenia characteristically becomes more obvious on sustained up-gaze.

Clinical pictures

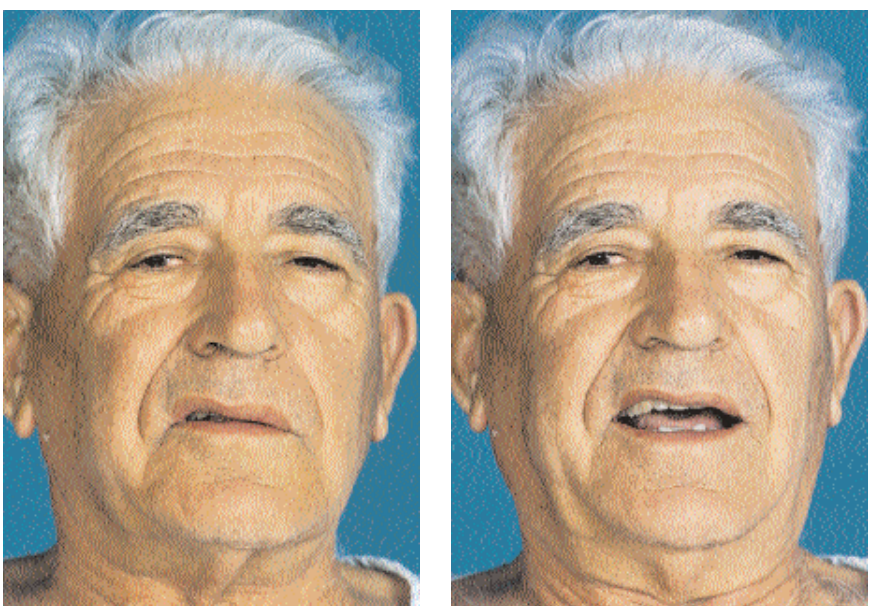
Myopathic facial weakness

Myopathies that affect the face frequently produce a 'horizontal smile' and bilateral ptosis (see Figure 3) caused by weakness of the muscles that elevate the angles of the mouth and eyelids, respectively. In myasthenia gravis, a neuromuscular junction disorder, ptosis becomes more noticeable when the patient is asked to look upward at a target for 60 to 90 seconds (fatiguable weakness).

The smile of patients with myopathic facial weakness may have a distinctive snarling quality. The condition may be accompanied by weakness involving other muscle groups such as the neck flexors, shoulder girdle, pharynx or palate. Weakness of the palate leads to dysphagia or nasal regurgitation of fluids.

Impaired emotional expression

Expression of emotion requires a network of brain regions, including the cerebral cortex, basal ganglia and limbic system. These regions all feed into the facial nucleus via distinct but normally interdependent pathways (see Figure A).



Figures 4a (above left) and b (above right). A 68-year-old man who suffered an ischaemic stroke in the right frontoparietal region. a. Left facial weakness in an upper motor neuron pattern, affecting chiefly the lower face, is most evident when he is asked to show his teeth in a voluntary smile. b. When telling a joke, the 'emotional' smile is more symmetrical.

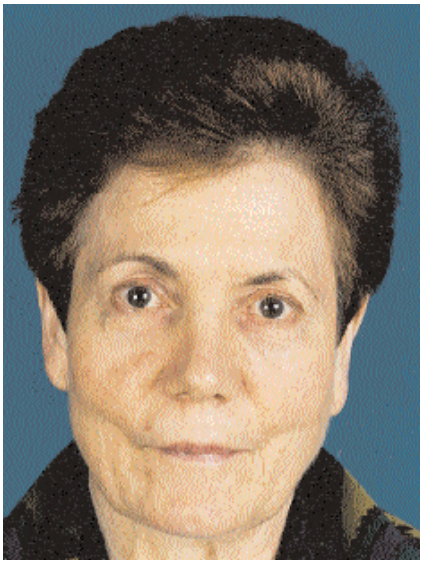


Figure 5. A 65-year-old woman with multiple system atrophy, an uncommon basal ganglia disorder. Her facial expression is impassive, with a characteristic unblinking 'reptilian' stare. Loss of emotional and automatic facial movements is a hallmark of basal ganglia disorders.

We can all distinguish genuine emotional expressions from posed ones of social display. It is sometimes observed that spontaneous, emotional smiling (such as that in response to a humourous remark) is preserved in an upper motor neuron lesion which produces asymmetry of a posed smile (Figures 4a and b).

Automatic facial responses (such as frowning with concentration), which are not associated with strong emotional arousal, may result from the action of the basal ganglia on the parts of the facial nuclei that innervate chiefly the upper face.

Lesions in these pathways account for the masked or 'staring' facies of Parkinson's disease, other basal ganglia disorders (Figure 5), and some forms of frontal lobe disease.

Pathological affect

The effects of loss of normal cortical controls over emotional expression are in motor neuron disease, diffuse cerebral atherosclerosis and multiple sclerosis. The emotional displays are externally indistinguishable from the normal motor patterns. In most cases, the provocation is trivial or nonspecific and the inner emotional state is neutral or even at odds with the outward display.

Pathological affect should be distinguished from other situations in which emotional lability and euphoria result from diffuse organic brain pathology (such as the fatuous affect of some forms of frontal lobe disease) or the pathological emotions of psychosis and drug intoxications.

Pathological affect is usually the consequence of bilateral lesions of the descending corticofacial pathways. Some therapeutic success has been claimed with tricyclic antidepressants.

Conclusions

In most disorders of facial movement, recognition of a small number of clinical patterns permits accurate localisation of the site of pathology. The time course and other features of the history often hold clues to the disease process producing the lesion.

The general practitioner has a key role in identifying patients who do not conform to well recognised or benign patterns of facial weakness and need specialist referral. MT

Next month, Part 2 of this article will discuss disorders that result in excessive facial movement.

Consultant's comment

The authors have produced an excellent two-part article discussing the afflictions which can, surprisingly commonly, affect the facial nerve or facial movement. As noted, there is a very wide variety of causes of these disorders, some transitory, some eminently treatable, and some very sinister. I think every GP should seize the opportunity to read the well-written descriptions, and to keep at hand the thoughtfully laid out tables. Among the causes of unilateral lower motor neuron lesions the acoustic neuroma is noted, along with the association with unilateral deafness. The deafness may in fact precede any facial nerve disturbance by months or years, so it is vital to keep this tumour in mind when investigating unilateral deafness, and get an early CT scan. Acoustic neuromas are curable with preservation of facial nerve function, and even occasionally of hearing, if detected when still small (less than 1 cm in diameter).

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