Geniculate neuralgia

Geniculate neuralgia is a craniofacial pain syndrome associated with the nervus intermedius.

General information

Geniculate neuralgia (GeN) AKA Hunt's neuralgia AKA nervus intermedius neuralgia: a very rare neural- gia affecting the nervus intermedius (the somatic sensory branch of the facial nerve primarily innervat- ing mechanoreceptors of the hair follicles on the inner surface of the pinna and deep mechanoreceptors of nasal and buccal cavities and chemoreceptors in the taste buds on the anterior 2/3 of the tongue). Symptoms: unilateral paroxysmal otalgia (lancinating pain experienced deep within the ear, often described as an "ice pick in the ear") radiating to the auricle, with occasional burning sensations around the ipsilateral eye and cheek, and prosopalgia (pain referred to deep facial structures, includ- ing orbit, posterior nasal, and palatal regions). During pain attacks, some patients have: salivation, bitter taste, tinnitus, or vertigo. GeN occasionally has cutaneous trigger points in the anterior EAC and tragus, and pain may also be triggered by cold, noise, or swallowing. Work-up includes neuro-otologic evaluation with audiometry and ENG. Some patients may require imaging (MRI or high-resolution CT) and angio (to R/O aneurysm).

Variants

Tic convulsif (AKA convulsive tic): Geniculate neuralgia combined with hemifacial spasm, usually due to neurovascular compression of both the sensory and motor roots of the facial nerve [1] , most often by AICA. First described by Cushing in 1920. GeN may be associated with herpetic infections of the geniculate ganglion (AKA herpetic ganglionitis, AKA Ramsay Hunt syndrome (RHS)) in which case herpetic lesions appear on the pinna, in EAC, and possibly on TM. May include facial palsy, decreased auditory acuity, tinnitus or vertigo. Unlike idiopathic GeN, RHS is more chronic and less paroxysmal, tends to remit with time, and is usually refractory to carbamazepine. Idiopathic GeN tends to be more painful than RHS, and does not remit spontaneously.

Epidemiology

Fewer than 150 reported cases were published in English between 1932 and 2012 [2].

Etiology

The etiology of the condition remains unknown [3].

Symptoms

Unilateral paroxysmal otalgia radiating to the auricle.

Ocassional burning sensations around the ipsilateral eye and cheek, and prosopalgia.

The pain may also be of gradual onset and of a dull, persistent nature, with occasional sharp, stabbing pain like an electric shock, deep in the ear [4].

Some people have reported additional symptoms during pain attacks:

Salivation

Bitter taste

Tinnitus

Vertigo

Diagnosis

The clinical presentation varies. Non-neuralgic causes of otalgia should always be excluded by a thorough clinical examination, audiological assessment and radiological investigations before making a diagnosis of geniculate neuralgia [5].

Differential diagnosis

Due to the close anatomical proximity, temporomandibular joint (TMJ) pathologies should be included in the differential diagnosis.

Easily confused with trigeminal neuralgia and glossopharyngeal neuralgia. However, nerves intermedius has its characteristic clinical syndroms to be diagnosed.

see Ramsay Hunt syndrome.

Treatment

see Geniculate neuralgia treatment.

Case series

see Geniculate neuralgia case series

Case reports

A 70-year-old woman who had deep-ear pain for more than 4 years and was diagnosed with trigeminal neuralgia and treated with carbamazepine without relief. Magnetic resonance tomographic angiography revealed no neurovascular conflict with the trigeminal nerve, whereas the anterior

inferior cerebellar artery (AICA) was close to the VII/VIII complex. We performed left-sided suboccipital retrosigmoid craniotomy. Surgical exploration under endoscopy clearly showed that the nervus intermedius was compressed by the AICA from behind. The ear pain was completely relieved immediately after nervus intermedius sectioning. The intraoperative findings and postoperative results confirmed that the compression of the nervus intermedius by the AICA caused the otalgia. A patient's specific pain, combined with preoperative imaging examination, is useful in the diagnosis of NIN. Neuroendoscopy has the advantages of enabling a clear field of view and close observation, thus aiding in the identification and accurate cutting of the nervus intermedius during the operation [6].

2014

A case illustration was presented that demonstrates the novel brainstem functional imaging findings for geniculate neuralgia. A 39-year-old man presented with a history of left “deep” ear pain within his ear canal. He noted occasional pain on the left side of his face around the ear. He had been treated with neuropathic pain medications without relief. His wife described suicidal ideations discussed by her husband because of the intense pain.

The patient's neurologic examination was normal, and otolaryngologic consultation revealed no underlying structural disorder. Anatomic imaging revealed a tortuous vertebral artery-posterior inferior cerebellar artery complex with the posterior inferior cerebellar artery loop impinging on the root entry zone of the nervus intermedius-vestibulocochlear nerve complex and just inferior to the root entry zone of the facial nerve and a small anterior inferior cerebellar artery loop interposed between the cranial nerve VII-VIII complex and the hypoglossal and glossopharyngeal nerves. A left-sided retromastoid craniotomy was performed, and the nervus intermedius was transected. An arterial loop in contact with the lower cranial nerves at the level of the brainstem was mobilized with a polytetrafluoroethylene implant.

The patient indicated complete relief of his preoperative pain after surgery. He has remained pain-free with intact hearing and balance [7].

2007

Figueiredo et al., present a case report of a female patient who was successfully managed with pharmacological treatment [8].

1984

A patient had combined otalgia and intractable unilateral facial spasm, relieved by microsurgical vascular decompression of the seventh and eighth cranial nerve complex in the cerebellopontine angle without section of the intermediate nerve. A dolicho-ectatic anterior inferior cerebellar artery compressed the seventh and eighth cranial nerves complex, suggesting that vascular compression of the intermediate nerve or of the sensory portion of the facial nerve may cause geniculate neuralgia. “Tic convulsif” seems to be a combination of geniculate neuralgia and hemifacial spasm. This combination could be due to vascular compression of the sensory and motor components of the facial nerve at their junction with the brainstem [9].

1)

Yeh HS, Tew JM. Tic Convulsif, the Combination of Geniculate Neuralgia and Hemifacial Spasm Relieved by Vascular Decompression. Neurology. 1984; 34:682–683
[2] [3] [5]
Tang IP, Freeman SR, Kontorinis G, Tang MY, Rutherford SA, King AT, Lloyd SK. Geniculate neuralgia: a systematic review. J Laryngol Otol. 2014 May;128(5):394-9. doi: 10.1017/S0022215114000802. Review. PubMed PMID: 24819337.
[4]
Pulec JL. Geniculate neuralgia: diagnosis and surgical management. Laryngoscope. 1976 Jul;86(7):955-64. PubMed PMID: 933690.
[6]
Song Z, Chen J, Shen J, Jia Z, Wang Q, Jiang S, Xu X, Shi W. Endoscopy during neurotomy of the nervus intermedius for nervus intermedius neuralgia: a case report. Ann Transl Med. 2021 Jan;9(2):179. doi: 10.21037/atm-20-5951. PMID: 33569481; PMCID: PMC7867896.
[7]
Tubbs RS, Mosier KM, Cohen-Gadol AA. Geniculate neuralgia: clinical, radiologic, and intraoperative correlates. World Neurosurg. 2013 Dec;80(6):e353-7. doi: 10.1016/j.wneu.2012.11.053. PubMed PMID: 23178920.
[8]
Figueiredo R, Vazquez-Delgado E, Okeson JP, Gay-Escoda C. Nervus intermedius neuralgia: a case report. Cranio. 2007 Jul;25(3):213-7. Review. PubMed PMID: 17696039.
[9]
Yeh HS, Tew JM Jr. Tic convulsif, the combination of geniculate neuralgia and hemifacial spasm relieved by vascular decompression. Neurology. 1984 May;34(5):682-3. PubMed PMID: 6538661.