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Research Paper

Glossopharyngeal Neuralgia By Neurovascular Compression with Posterior Inferior Cerebellar Artery (Pica)- A Case Report With Review

MEDICAL SCIENCE

KEYWORDS : Glossopharyngeal neuralgia (GPN), dolichoectesia, posterior inferior cerebellar artery (PICA), MRI

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ABSTRACT

Glossopharyngeal neuralgia (GPN) is a rare disorder with incidence of 0.7 in per 10,000 people. It usually begins after the age of 40 and commonly in men. It is characterized by paroxysm of unilateral severe lancinating pain in the tongue, throat, tonsil and may radiate to the ear. The pain is acute in onset and sharp, stabbing or burning in quality and may last for few second to few minutes and can be provoked by talking, chewing, swallowing, coughing and yawning. Glossopharyngeal neuralgia is believed to be caused by irritation of the the glossopharyngeal nerve. The causes of GPN are diverse and are usually divided into two groups, idiopathic and secondary causes such as neoplasm, inflammatory diseases, infections, chiari I malformation, infaction and neurovascular compression. In our case GPN found to be caused by compression of left glossopharyngeal nerve by left PICA.

Introduction

Glossopharyngeal neuralgia is a rare entity, it is characterized by paroxysm of unilateral severe lancinating pain in the tongue, throat, tonsil and may radiate to the ear. The distribution of the pain in GPN can be otitic or can be oropharyngeal. In otitic type pain is in and around the ear, mastoid region while in oropharyngeal type pain in the side or back wall of the pharynx, posterior third of the tongue, fauces, tonsil, or soft palate similar to our patient. Oropharyngeal pain radiates to the ear or adjacent regions in the majority of cases like our patient while otitic pain does not often radiate. The pain is acute in onset and sharp, stabbing or burning in quality and may last for few second to few minutes and can be provoked by talking, chewing, swallowing, coughing and yawning. Some may experience neuralgia as clicking, scratching and foreign body sensation, and rarely there can be palatal myoclonous, syncope and even cardiac arrest. Glossopharyngeal neuralgia is believed to be caused by irritation of the ninth cranial nerve, called the glossopharyngeal nerve. The causes of GPN are diverse and are usually divided into two groups. Secondary underlying causes include such as neoplasm, inflammatory diseases such as multiple sclerosis(MS), infections, chiari I malformation, infaction and neurovascular compression. Patients with GPN and without an identifiable underlying pathological process are referred to as having essential or idiopathic GPN. In most of the cases the cause of irritation could not be localized.

In case of nerurovascular compression the most common offending vessel has been reported as posterior inferior cerebellar artery (PICA) followed by vertebral, anterior inferior cerebellar artery (AICA) and other.

Case report

A 85 year female presented with sudden recurrent attacks of pain in her thought while chewing and swallowing food since last 18 months. This pain was of sharp, jabbing, electric, shock like and whenever occur, located deep in the left half of the throat, some time extends upto the same side ear. Patient was non diabetic, normotensive, non smoker, not having any previoug history stroke. Her blood pressure was 134/88mmHg in supine, pulse rate was 88/min, weight 56kg, BMI was 23.4kg/m^2. She is not having any history of weight loss or pain anywhere in the body. On examination of the neck neither any swelling nor lymphadenopathy was found. Oral examination teeth, buccal and gingival mucosa, tongue, tonsils, palate, posterior pharyngeal wall were found to normal. There was no history fever and any kind of surgery or irradiation in past. So we planned for the imaging study and obtained a MRI brain and soft tissue neck to ensure that there was no evidence for a tumor or other lesion, and then we found that Left side vertebral artery is grossly dolichoectatic and left side PICA is seen compressing up on the left glossopharyngeal nerve while other tissue were normal.

Discussion

Glossopharyngeal Neuralgia is like Trigeminal Neuralgia but not as common.The incidence of GPN is about 0.7 per 100,000 people in the general population [1]. Glossopharyngeal neuralgia usually begins after age 40 and occurs more often in men. The distribution of the pain in GPN can be otitic or can be oropharyngeal. In otitic type pain is in and around the ear, mastoid region while in oropharyngeal type pain in the side or back wall of the pharynx, posterior third of the tongue, fauces, tonsil, or soft palate similar to our patient. Oropharyngeal pain radiates to the ear or adjacent regions in the majority of cases like our patient while otitic pain does not often radiate.

Most often the onset is sudden and spontaneous; however a gradual onset of an increasingly severe pain also occurs. The attacks tend to occur with a frequency of one to five per year. The attacks usually last from several weeks to one to two months with pain-free days during the attack period. The duration of the neuralgic pain is usually from seconds to minutes (up to 20 minutes). The number of these neuralgic pains in a 24-hour period commonly averages between five to thirty(up to one hundred) and they occur during day and night. The severity of the pain is also extremely variable (from mild to severe). Mild attacks occur and cause minor inconvenience. Usually patients are stable between the attacks, but some of them have mild, episodic 'sticking sensations' in the same area as the pain. The most commonly observed precipitating factor is swallowing (saliva, solid foods and liquids), which was present in our patient. Chewing and talking have also been described as triggers, as in our patient. Infrequent causes are yawning, movements of the head, straining and mouth movements. Certain foods, independent of the

act of swallowing, may precipitate an attack (bitter, spicy, salty), but never sweet [2].

Other non-painful clinical manifestations include sensations of a swollen soft palate, headache, and hyperesthesia, which may occur for several hours, usually in a secondary field, after the pain has subsided. Other less frequent presentations of GPN consist of burning sensations in the muscles of the neck, difficulty in swallowing liquids and solids, excessive secretion of saliva, bilateral tinnitus, bilateral lacrimation, flushing and sweating, irritation in the throat and excessive coughing, dizzy attacks, objective vertigo and unsteadiness, bradycardia and even cardiac arrest with syncope [3].

The causes of GPN are diverse and are usually divided into two groups. Secondary underlying causes such as neoplasm or inflammatory diseases, such as MS, affect the glossopharyngeal nerve at different levels of its path at brainstem structures, cisternal branches, jugular foramen, and extracranial divisions. Patients with GPN and without an identifiable underlying pathological process are referred to as having essential or idiopathic GPN. Idiopathic GPN according to some authors is caused by compression of cisternal part of the glossopharyngeal nerve by intracranial vessels [4]. A three-dimensional MRI scan of the brain revealed that contact or contiguity between blood vessels and neural tissue is not visibly different on the symptomatic and asymptomatic sides, thus discounting any consistent causal relationship to pain [5]. In our patient, we demonstrated a definitive contact between the PICA and glossopharyngeal nerve that could explain our patient's symptomatology. Diagnostic investigation for GPN should exclude MS and local compression, particularly by cancerous lesions, which require more aggressive treatments. Vascular compression is a common and potentially treatable cause of GPN but does not account for all previously designated idiopathic cases. The proposed mechanisms of irritation of the glossopharyngeal nerve and the resultant painful paroxysms include hyperexcitability of the glossopharyngeal nerve and ephaptic conduction; however, none have been fully explored. Based on current hypotheses, it is suggested that compression of the glossopharyngeal nerve at the nerve root entry zone results in demyelination and ephaptic transmission among the glossopharyngeal nerve axons; another theory suggests that vascular compression of a nerve induces repetitive activation of primary afferents in the nerve and leads to hyperactivity and hyperexcitability in the central neurons; the last implies activation of *N*-methyl- D-aspartic acid receptors in the compressed nerve [6].

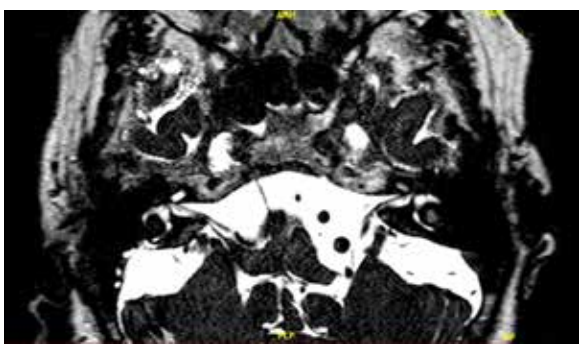
The best diagnostic test is the topical application of cocaine or viscous xylocaine to the throat and tonsilar fossa on the affected side, which should provide temporary pain relief and can be used to allow the patient to eat as well as to confirm the diagnosis..

Medical treatment of GPN with carbamazepine or gabapentin is effective in the majority of patients. Our patient responded well to carbamazepine at a moderate dose. In cases refractory to medical therapy surgical methods could be applied. Surgical methods include destruction of petrous ganglion (radiofrequency percutaneous rhizothomy, Gamma Knife® stereotactic radiosurgery), extracranial (neurotomy) or intracranial (rhizotomy) resection of glossopharyngeal nerve [7], separation of the vessel from the nerve (MVD) [8], destruction of glossopharyngeal nerve ascending fibers (tractotomy) [9], motor cortex stimulation, thalamic and other deep brain stimulation, and centrally administered opioids. More recently, endoscopy has been employed as the sole modality in glossopharyngeal nerve decompression [10].

Conclusion

In conclusion, glossopharyngeal neuralgia due to neurovascular compression is very rare condition. PICA is one of among most common offending vessels. In our case left sided PICA is compressing the left glossopharyngeal nerve. MRI is very beneficial in case of glossopharyngeal neuralgia and an offending artery that can be localized by MR angiography. Otherwise most of the cases responded well with pharmacotherapy like in our case but in some refractory cases surgery can be beneficial.

IMAGE



T2W MRI of neck showing left PICA compressing the G.N. and left vertebral artery is dolichoectatic

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