KJP The Korean Journal of Pain
| Case Report |

Korean J Pain 2010 September; Vol. 23, No. 3: 215-218
pISSN 2005-9159 eISSN 2093-0569
DOI : 10.3344/kjp.2010.23.3.215

Severe Pain Attack Associated with Neurocardiogenic Syncope Induced by Glossopharyngeal Neuralgia: Successful Treatment with Carbamazepine and a Permanent Pacemaker
-A Case Report-

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Glossopharyneal neuralgia (GPN) is generally considered to be a pain disease. However, it can be also be a life-threatening cardiac cause of syncope. Neuralgia in the throat and neck can trigger severe bradycardia up to the point of asystole, which can progress to cardiac syncope with or without seizures. A 65 year-old male patient diagnosed with glossopharyngeal neuralgia complained of severe paroxysmal pain in his right chin and ear followed by bradycardia, aystole and syncope. We report a case successfully treated with a permanent pacemaker and carbamazepine in a patient with GPN who had syncopal attacks preceded by paroxysms of pain. (Korean J Pain 2010; 23: 215-218)

Key Words:
carbamazepine, glossopharyngeal neuralgia, permanent pacemaker, syncope.

Glossopharyngeal neuralgia (GPN) is an uncommon neuropathic condition characterized by paroxysms of unilateral and severe stabbing pain felt in the ear, base of the tongue, tonsillar fossa or beneath the angle of the jaw. It is commonly incited by swallowing, talking or coughing, which usually lasts from seconds to minutes, and may remit and relapse in the manner of trigeminal neuralgia (TN). It is not uncommon that some clinicians may misdiagnose GPN as TN.

Occasionally, GPN can be associated with cardiac syncope, which in most instances is caused by bradycardia or asystole [1,2]. This syndrome may be referred to as vagoglossopharyngeal neuralgia when the cardiac symptoms accompany pain attacks [1].

We report here a case of GPN, initially diagnosed as TN and associated with severe bradycardia, asystole and subsequent syncope successfully treated by a permanent pacemaker and carbamazepine.

Received May 31, 2010. Revised July 23, 2010. Accepted July 23, 2010.
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CASE REPORT

A 65-year-old man with no medical history was transferred from another hospital because of pain in the right side of chin and ear. He complained of frequent paroxysms of severe stabbing pain in the right side of his mandible radiating to the right ear. His pain either occurred spontaneously or was triggered by chewing and tooth brushing and lasted few seconds in each pain attack. He had experienced the pain since 5 months ago. The patient was diagnosed as TN at another hospital and was prescribed carbamazepine (200 mg a day) with only partial relief of the pain. He had a pain-free period for about 1 month before visiting our clinic and stopped medication on his own decision. In the last few days prior to the admission, his condition had got worse in terms of increase of frequency and severity of pain. He visited our clinic because of intractable pain even with medications.

The routine laboratory tests and electrocardiogram were normal. Neurological and nasopharyngeal assessments were normal findings. Computed tomography of the neck and magnetic resonance imaging (MRI) of the brain revealed also normal. His symptoms were fulfilled in the diagnostic criteria of TN [3].

His pain paroxysms had occurred four or five times daily with 100 of visual analogue scale (VAS, 0 is no pain and 100 is imaginary the worst pain) at the first visit. The duration of pain attacks was measured in a few seconds. He underwent mandibular nerve block with alcohol after the test block with 1% of mepivacaine. He still complained of right ear pain even though right V3 innervated area was anesthetized and pain free after the V3 alcohol block. Ten minutes after the procedure, paroxysmal pain in right ear developed and he experienced faintness and got loss of consciousness with convulsive movement for about 1 minute. He was consulted with a neurologist to exclude seizure disorders. Electroencephalogram showed no abnormalities. Brain MRI was not able to be performed due to convulsive movement during the pain attack.

He was consulted to a cardiologist. His echocardiogram was normal, however, bradycardia followed by asystole during pain attacks was revealed in the continuous electrocardiography. The duration of bradycardia and asystole was in proportion to duration of pain attack. According to a careful asking about the pain characters following the V3 block with alcohol, he presented that his pain occurred in the soft palate, uvula and throat and was triggered by swallowing or mechanical stimulation of the right side of the pharynx. Bradycardia and asystole accompanied with sometimes seizure-like activity occurred always with pain attack. Based on his altered pain characters and cardiac symptoms with no specific etiology originated from heart, he was diagnosed of GPN associated with asystole.

Carbamazepine (100 mg twice a day) was started for control of pain as well as asystole. Regardless of increasing dose of carbamazepine up to 400 mg a day, severe bradycarida, asystole and syncope preceded by paroxysmal pain continued four to five times in one hour. A temporary cardiac pacemaker was implanted via left subclavian vein by a cardiologist to prevent cerebral ischemia during asystole. Although syncope and seizure-like activity disappeared after the insertion of a temporary pacemaker, intensity or frequency of pain in the throat and ear was unchanged. Carbamazepine increased up to 600 mg a day to reach pain control for 4 days after the implantation of temporary cardiac pacemaker. On the 5 day of hospitalization, he underwent implantation of a permanent pacemaker to prevent bradycardia and asystole by a cardiologist. He was achieved pain free and no cardiac symptoms after the implantation of permanent pacemaker with daily 600 mg of carbamazepine and has maintained symptom free condition for 4 months of follow-up.

DISCUSSION

GPN is an uncommon form of facial pain (0.2 to 1.3% of the facial pain) that occurs approximately one hundred times less frequently than TN. It was first described by Weisenberg in 1910 in a patient with tumor affecting cerebellopontine angle [4]. Most common cause of idiopathic GPN is vascular compression of the glossopharyngeal nerve. The other secondary causes of GPN are following; cerebellopontine angle tumor, carcinoma of the laryngeal and nasopharyngeal tumors, parapharyngeal abscess, multiple sclerosis, trauma, direct carotid puncture, Paget's disease, calcified sylohyoid ligament, and Chiari I malformation [1,2,5].

It is generally affected in adults, especially over 50 years of age [1]. There is no consensus regarding which sex is most affected [6]. Pain characters are almost similar to TN, however, pain originates in the ear, posterior phar-

ynx, tonsillar area and base of the tongue innervated by glossopharyngeal nerve. Even though majority of GPN patients report that swallowing is a prominent trigger factor, some of TN patients also have it as a pain-provocating factor. Because of the similar pain characters between GPN and TN, especially in the V3 division, it could be misdiagnosis GPN as TN. Concurrent ipsilateral TN and GPN is rare, representing 10.0% to 46.7% of GPN cases but only 0.3% to 0.5% of TN cases [7].

Detailed history taking may help to distinguish GPN from TN. The pain is triggered by pharyngeal movement such as coughing, gargling and swallowing, especially cold liquids, as well as by touching in the above mentioned zones. While in TN, the pain is confined to the face, and is triggered by facial movement, such as chewing and speaking. In our patient he visited our clinic with known TN and presented TN symptom at the first visit, however, he could express the GPN component of pain when TN component of pain disappeared after the V3 alcohol block. We might conclude that our patient had TN with GPN.

The association between GPN and syncope is very rare phenomenon and could be life-threatening. It was reported that 217 patients diagnosed for GPN and only 4 were found having associated syncope [1].

Mechanisms of cardiac and cerebral manifestations produced by GPN are not completely understood. Postulating mechanism is originated from the idea of a close connection between the glossopharyngeal and vagus nerves with circulatory system. It has been suggested that intense afferent impulses from the sensory fibers of the glossopharyngeal nerve may stimulate the dorsal motor nucleus of the vagus nerve either by way of central collateral pathways [8] or through an "artificial synapse" along the peripheral course of the glossopharyngeal nerve as it travels with the nerve of Hering [9]. In neuralgia pain condition like GPN, extremely severe pain could activate the vagoglossopharyngeal reflex resulting in bradycardia, hypotension and syncope.

The basic goal of treatment of GPN with cardiac manifestations should be focused on control of the pain, which could be a main cause of bradycardia and syncope. There are two options to treat pain; medical and surgical treatments [10]. First choice of medical treatment is carbamazepine and other medical trial is gabapentin although theoretically any membrane stabilizer could be used. The problem with using carbamazepine is the possibility of tachyphyaxis due to need of long-term use. In our case carbamazepine was effective at a moderate dosage (600 mg/day) in abolishing paroxysmal pain. Although percutaneous radiofrequency thermocoagulation through oval foramen has been tried, it carries the risk of injury to the whole vagus nerve [11]. Microvascular decompression might be considered as an effective treatment for GPN. Kondo reported that immediate complete relief was achieved in 67 to 79% of patients, and partial relief in 10 to 25% of patients obtained, while long-term complete relief was obtained in 58 to 76% and partial relief in 15 to 18% of patients [12]. However, manipulation of lower cranial nerves can be associated with morbidity; dysphagia, hoarseness, facial paresis, hearing disturbance associated 7th, 8th, 9th and 10th caranial nerve disturbances, and cerebrospinal fluid leakage [12].

A temporary pacemaker should be used for emergency management of syncopal attacks caused by bradycardia and asystole originated from GPN like our patient until plasma concentration of carbamazepine reaches the therapeutic range. A temporary transvenous cardiac pacemaker implantation was described first by Khero and Mullins [13] in 1971 to treat the reflex cardiac syncope while waiting for surgical section of the nerve.

Regarding permanent pacemaker implantation, the available literatures are quite controversial [1,14], because spontaneous recovery in GPN is not uncommon and relapses may occur. However, once severe bradycardia and asystole relapse again in unexpected time, the patient might be at risk of a fatal outcome. It should be individualized to determine to implant a permanent pacemaker in patients with GPN associated with neurocardiogenic syncope. In our case, a permanent pacemaker underwent to prevent heart and brain damage with the reasons of that every pain attack leaded to asystole and relatively old patient's age.

In conclusion, a temporary pacemaker combined with medical treatment should consider as an initial treatment modality in patients with GPN associated with asystole and syncope. Furthermore, if the disorder is longstanding and severe, a permanent pacemaker combined with medical treatment could be a safe treatment option.

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