Glossopharyngeal neuralgia with cardiac syncope treated by glossopharyngeal rhizotomy and microvascular decompression

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A glossopharyngeal neuralgia case with cardiac asystole is presented. The sinus mode dysfunction and subsequent syncope with pain appears to be the most important life-threatening symptom in the late period of the disease. Because of cardiac symptoms induced by intense vagal stimulation, this case was considered to be vagoglossopharyngeal neuralgia. Several medical and surgical alternatives have been proposed for its treatment. In the presented case the cause of disease was compression of lower cranial nerves in the right cerebello-pontine angle (CPA) by the vertebro-basilar arterial system. Carbamazepine and pacing were determined ineffective, so the patient was treated surgically by cutting the glossopharyngeal nerve and by decompression of the vagal nerve at the CPA. The sinus arrest and paroxysmal pain attacks disappeared and the patient’s life returned to normal.

Introduction

Glossopharyngeal neuralgia (GN) is an uncommon craniofacial syndrome characterized by paroxysms of unilateral and severe stabbing pain occurring in the distribution of the nerve. The pain may be spontaneous or precipitated by swallowing, chewing, yawning, coughing or talking[1]. Occasionally, GN can be associated with cardiac syncope, which in most instances is caused by asystole or bradycardia and, possibly, by a vasodepressor reaction[2–5]. This syndrome may be referred to as vagoglossopharyngeal neuralgia when the cardiac symptoms accompany pain attacks[5,9,11]. Several medical and surgical alternatives have been proposed for the treatment of GN associated with syncope[1,5,8,9,13]. Temporary and permanent pacemakers have been used to prevent attacks of syncope with different results[2–6]. Surgical treatment has also evolved over time with very good results reported[1,8,11,12,13]. Here, we present a case of GN associated with cardiac asystole and subsequent syncope successfully treated by glossopharyngeal rhizotomy and microvascular decompression.

Case report

A 51-year-old woman reported a 2-year history of paroxysmal pain attacks originating from the right lower jaw and disseminating to the right ear region when eating and swallowing. The duration of pain attacks was measured in seconds. These pain paroxysms had occurred two or three times daily at the initial stage of the disease. The pain attacks disappeared for a 2- to 3-month period. Six months ago, the patient was admitted to the Department of Neurosurgery with complaints of paroxysmal glossopharyngeal pain and presyncopal attacks, and was hospitalized. During her hospitalization, carbamazepine was stopped but...
paroxysmal pain attacks ceased and no cardiac arrhythmia was observed so the patient was discharged. Carbamazepine (600 mg daily) was started in order to prevent presyncope and pain attacks. After a 2-month period, paroxysmal glossopharyngeal pain attacks gradually recurred and increased. In the 2 weeks prior to her admission for placement of temporary pacing, the patient complained of presyncopal episodes during her pain attacks. She was admitted to the hospital with severe paroxysmal pain attacks in addition to syncopal episodes. Telemetric recordings taken during the attacks showed bradycardia and sinus arrest (maximum 6·6 s) (Fig. 1). General physical examination, and cardiovascular and neurological examination findings were normal. Massage of the carotid sinus did not cause bradycardia. The pain and syncopal attacks were not completely controlled by carbamazepine at a daily dose of 1200 mg.

Magnetic resonance images and magnetic resonance angiography (MRA) (Fig. 2) demonstrated that both of the vertebral arteries were abnormally tortuous and fused in the right CPA, compressing the glossopharyngeal and vagal nerves. For abbreviations see text.

Figure 1  Sinus arrest lasting 6·6 s with pain and syncopal attack.

Figure 2  MRA: Both of the vertebral arteries (V.A.) were abnormally tortuous and fused in the right CPA, compressing the glossopharyngeal and vagal nerves. For abbreviations see text.
VVI pacing was also used during surgery to prevent bradycardia.

During the operation the right CPA was approached. Both of the vertebral arteries were found just under the lower cranial nerves (IXth, Xth, XIth). Basilar bifurcation emerged from the right CPA, compressing the IXth, Xth, and XIth cranial nerves and brain stem. The IXth cranial nerve (glossopharyngeal) was sectioned (Fig. 3). Vertebro-basilar system was gently dissected and vagal rootlets and brain stem were decompressed by a piece of muscle. After the operation, pain was completely relieved, and carbamazepine was stopped. No episode of sinus arrest was found on a 24-h Holter monitor. During the follow-up period of 3 months, no sinus arrest or pain has occurred.

**Discussion**

Glossopharyngeal neuralgia is an uncommon form of facial pain. It usually occurs without any obvious cause. Most of these cases of ‘idiopathic’ GN are caused by vascular compression of the glossopharyngeal nerve\[^1,6\]. The other secondary causes of GN are Eagle’s syndrome, cerebellopontine angle tumors, parapharyngeal space lesions, multiple sclerosis, arachnoiditis, posterior fossa arteriovenous malformation, direct carotid puncture, metastatic head and neck tumors, and Chiari I malformation\[^1,6,7,10,13,14\]. GN is occasionally associated with cardiac syncope. In the large Mayo Clinic series of 217 cases of GN, four patients with syncope were reported\[^9\]. Various reports and case studies demonstrating this relationship have been published in the literature\[^2,6,9,11\].

The sinus dysfunction and subsequent syncope with pain is probably due to the spread of afferent impulses from the trigger zones to the dorsal motor nucleus of the vagus, causing intense vagal stimulation. In the presented case, we concluded that the hyperstimulation of the glossopharyngeal and vagal nerves was induced by compression by the vertebro-basilar arteries (Fig. 3). Because of these relations and symptomatology, we named this syndrome as vagoglossopharyngeal neuralgia.

Carbamazepine can prevent paroxysms of pain and associated cardiovascular manifestations\[^9,15\]. In our case, carbamazepine was not effective at a dosage of 1200 mg/day.

A temporary transvenous cardiac demand pacemaker was proposed by Khero and Mullins in 1971 to prevent attacks of syncope\[^3\]. Jamshidi and Masroor\[^3\] and Alpert et al.\[^4\], in two other reports, successfully treated their patients with carbamazepine and permanent demand pacemakers because the patients also manifested hypersensitive carotid sinus syndrome. Johnston and Redding published the results of long-term treatment with a permanent pacemaker and carbamazepine in their patient without hypersensitive carotid sinus syndrome. Symptoms were controlled during a 10-year follow-up period\[^5\]. In our case, temporary VVI pacing controlled the syncopal attacks although episodes of neuralgia pain continued. Pacing does not affect pain but only controls the syncopal episodes and should be regarded as a palliative but potentially life-saving treatment. Furthermore, temporary and permanent pacemakers may not control the syncopal attacks in patients who have important vasodepressor components of their attacks\[^8,7,14\].

Surgery is the final step in treatment. The cause of disease in our case was compression of lower cranial nerves in the right CPA by the vertebro-basilar arterial system. Because the compressing arteries were the large arteries of the brain stem, the lower part of the CPA glossopharyngeal nerve was sectioned and enough space was obtained to dissect gently those large arteries. A piece of muscle was then placed between the arteries.
and brain stem. The Xth cranial nerve was completely decompressed. In classical literature, lower cranial nerves are usually compressed by the posterior inferior cerebellar artery (PICA), and its decompression is technically easy. Because of size of the artery (vertebrobasilar system) involvement in our case, this modified technique was chosen for decompression.

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References


