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Glossopharyngeal neuralgia associated with cardiac syncope: Two case reports and literature review



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ABSTRACT

INTRODUCTION: Vago-glossopharyngeal neuralgia is an unusual clinical syndrome characterized by paroxysms of sharp pain in the distribution of glossopharyngeal nerve. Rarely the condition is associated with arrhythmia and cardiac syncope, a phenomenon named vagoglossopharyngeal neuralgia.

PRESENTATION OF CASES: Here we present two patients with glossopharyngeal neuralgia associated with repetitive episodes of syncope referred from their primary care physician to neurosurgery department of a general hospital in Crete, Greece. The patients were successfully treated with microvascular decompression.

DISCUSSION: A literature review on pathogenesis, diagnosis and management is also performed.

CONCLUSIONS: Surgeons as well as primary care physicians have to be aware of this rare condition in order to prevent associated life-threatening complications and improve patient's clinical status with accurate therapy.

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1. Introduction

Glossopharyngeal neuralgia represents an unusual craniofacial clinical syndrome characterized by paroxysms of a stabbing pain in the distribution area of the glossopharyngeal nerve [1]. Rarely, in 2% of the cases, the condition is associated with cardiac arrhythmia and cardiac syncope [2]. In this case the syndrome is named vagoglossopharyngeal neuralgia [3]. We present 2 rare cases of glossopharyngeal neuralgia associated with repetitive episodes of syncope successfully managed with microvascular decompression. Taking this opportunity we also performed a literature review on pathogenesis, diagnosis and therapeutic alternatives of this entity.

2. Case 1

A 60 year old female was referred to our department by her general practitioner due to a 3 year history of paroxysmic sharp pain attacks located in the left side of the pharynx and the base of the tongue, with radiation to the ipsilateral cheek and ear. The paroxysms were usually triggered by swallowing and chewing,

and sometimes they were associated with loss of consciousness, 10–15 s of duration. The patient had already been referred to a cardiologist, an ENT specialist and a neurologist. The neurologist recommended treatment with carbamazepine at a final dose of 400 mg × 3, with no success in controlling the symptoms of pain, nor the episodes of syncope. The patient's physical/neurological exam revealed no abnormal findings and was admitted for further investigation. Electroencephalography, brain and neck magnetic resonance imaging as well as brain angiography were all normal. After vagoglossopharyngeal-associated syncope was diagnosed, surgical treatment was decided, because the patient has already received appropriate pharmacological treatment and her condition was still progressing. The patient was placed in the lateral decubitus position. A lateral suboccipital craniectomy was performed. Using the operating microscope, glossopharyngeal nerve and vagal rootlets were examined. Neurovascular compression was identified, with the presence of dense arachnoid adhesions between the nerves. Microvascular decompression consisted of surgical lysis of those adhesions. There were no postoperative complications and the patient did not suffer any episodes of pain or syncope postoperatively. No pharmacological treatment has been administered to the patient after surgery. During a 2 year follow-up the patient remained free of vagoglossopharyngeal neuralgia.

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3. Case 2

A 64-year old woman with a 2 year history of intense episodes of nidding pain to the left part of the pharynx, reflecting to the base of the tongue and the ipsilateral cheek, and syncopal episodes provoked by swallowing, was referred to our department. She had been previously referred to a cardiologist and an ENT specialist and was under antiarrhythmic treatment for the syncopal episodes, with no success in controlling them. Because of the progressively worsening symptoms of pain and syncope, treatment with carbamazepine was initiated and she was admitted for further investigation of a possible vagoglossopharyngeal neuralgia diagnosis. She was experiencing approximately two syncopal episodes per day, with a mean duration of 15 s each. During an episode of loss of consciousness that occurred within our clinic, the EKG revealed arrhythmia and premature atrial contractions. She underwent a series of tests, with CT and MRI of brain and neck not showing any abnormalities. Her angiography, on the other hand, showed a helicoid course of the left posterior inferior cerebellar artery (PICA), which was more dilated than the right one. Due to the initial substantial improvement with pharmacotherapy, the patient was released, only to be readmitted 5 days later because of a new syncopal episode. The patient was transferred to the operation room and was positioned in the lateral decubitus position. A lateral suboccipital craniectomy was performed. We exposed the vagal and glossopharyngeal nerves and after lysis of the arachnoid adhesions from their rootlets, we noticed that the left PICA was in contact with the glossopharyngeal nerve. We isolated the glossopharyngeal nerve from the PICA by placing a piece of Teflon sponge between them. At this point we decided to perform rhizotomy of the upper vagal rootlets, because they were close to the PICA and so, according to our assessment, there were better chances of controlling the syncopal episodes this way. During the postoperative course, the patient was free of pain, but on the first day she developed significant hypotension that was successfully treated with fluids, and suffered a slight loss of taste to the base of the tongue. She has been on follow-up by our department for 2 years and is free of symptoms, both pain and syncope.

3. Discussion

Glossopharyngeal neuralgia (GPN) is a rare facial pain syndrome, accounting for 0.2–1.3% of facial pain syndromes [3,4]. Its overall annual incidence is 0.8/100000 [5]. Approximately 10% of patients with GPN are misdiagnosed as trigeminal neuralgia [5,6]. This happens because both syndromes are manifested with facial pain. However in case of GPN pain is located unilateral and extends to the ear and throat [7]. Despite that it has been reported that bilateral pain may be present in less than 2% of the cases of GPN [6]. Rarely the syndrome is associated with episodes of cardiac syncope. In this case is called vagoglossopharyngeal neuralgia (VGPN) [2]. Syncopal episodes are manifested with bradycardia or asystole, hypotension and EKG changes that include various arrhythmias and premature atrial contractions, leading frequently to misdiagnosis of cardiac or cerebral episodes. GPN was first described by Weisenberg in 1910 [8] and in 1921 Harris announced that the syndrome may result to cardiac syncope [9]. GPN can be idiopathic or secondary to cerebellopontine tumors [8], vascular compression of the nerves in the posterior cranial fossa [10], laryngeal and rhinopharyngeal tumors [11], parapharyngeal abscesses [12], injuries [13], Paget's disease or skull base tumors [14] or even after carotid angiography [15]. In the majority of cases it occurs in the left side and in patients over 50 years old [10]. A preponderance in males has been also reported [3]. In regards to etiopathogenesis of cardiac syncope in the context of VGPN, a variety of mechanisms have been proposed. Some authors suggest that cardiac syncope is caused by irritation of the

nerve of Hering (carotid sinus nerve), which is an afferent branch of the glossopharyngeal, because of adhesions between the rootlets of the vagal and glossopharyngeal nerves [16,17]. Another view suggests that some visceral sensory fibers of the glossopharyngeal nerve might connect with fibers of the carotid sinus nerve (nerve of Hering), causing stimulation of the latter, therefore activation of the baroreceptors of the carotid sinus, resulting in bradycardia, asystole, premature atrial contractions or hypotension that can lead to syncope [16].

The diagnosis of idiopathic VGPN is based on a detailed medical history and a full neurological examination of the cranial nerves. It is also remarkable that convulsive syncope might be associated with glossopharyngeal neuralgia [18]. Seizures are in differential diagnosis with convulsive syncope because the latter has motor symptoms which may be confused with a tonic clonic seizure. Video EEG is useful to show that the motor symptoms are due to asystole and not to epileptic activity in the brain [18].

Interestingly Varrasi et al. (2011), reported an interesting case of VGPN in a 65 years old woman admitted with convulsive episodes preceded by stabbing pain extending from the left submandible zone to the neck and ipsilateral ear [18]. Activation of the dorsal motor nucleus of the vagus nerve by abnormally enhanced input from the glossopharyngeal nerve, via the tractus solitarius of the brainstem, has been reported to be one of the most accepted trigger mechanism for convulsive syncope in glossopharyngeal neuralgia [18]. In difficult cases, diagnosis can be established through application of local anesthetic on the affected area or glossopharyngeal anesthetic block with tetracaine; these tests are considered positive if the symptoms are temporarily relieved [6]. A variety of conservative and surgical treatment options have been suggested for the treatment of GPN, even in the case of associated syncope. Antiepileptic drugs are used to control the symptoms of idiopathic GPN, with carbamazepine being the main representative, but its efficacy in secondary GPN is strongly questioned [6]. For treating bradycardia caused by VGPN, isoproterenol and atropine have been used, with good results, but with no effect on the pain symptoms [19]. Surgical management is the treatment of choice when pharmaceutical treatment fails. Surgery consists of open intracranial rhizotomy of the glossopharyngeal nerve, with controversial results [1], and percutaneous rhizotomy of the glossopharyngeal nerve through the jugular foramen. Microvascular decompression (MVD) has been reported to be a successful technique especially when GPN is produced by compression of the nerve by a vessel [10]. There are a number of suggestions concerning the number of vagal nerve rootlets that should be sectioned. The vessel most often related with GPN is the posterior inferior cerebellar artery (PICA), and less often the vertebral artery, the anterior inferior cerebellar artery or any of the veins [10]. During surgery we noticed episodes of hypotension and arrhythmias, that didn't reoccur postoperatively. There was only one episode of postoperative hypotension in our second patient that was successfully treated with fluids. Although using a pacemaker can somehow control the syncopal episodes, it cannot relieve the patient from pain. For this reason as well due to the relatively low efficacy of drugs in secondary VGPN, we advocate surgical approach, especially in the presence of nerve adhesions or vascular compression. Through surgical treatment patients are relieved of both pain and syncope, whilst maintaining nerve functionality.

4. Conclusions

A high level of awareness upon this rare entity is required from surgeons as well as general practitioners. Since life threatening conditions rarely allow diagnostic revisions [20,21] timely diagnosis based on high vigilance is crucial in order to prevent associated

life-threatening complications and improve patient's clinical status with accurate therapy.

Conflict of interest

The authors declare that they have no conflict of interests.

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Ethical approval

No ethical approval was required.

Consent

Written informed consent was obtained from the patients for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Author contribution

AK and SM operated the patients. AK, DA and AH prepared the first draft of the manuscript.

MK and SM revised the manuscript for important intellectual content and technical details.

EKS provided useful suggestions on content and editing issues. All authors have read and approved the final manuscript.

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