

GLOSSOPHARYNGEAL NEURALGIA WITH SYNCOPE AS A SIGN OF NECK CANCER RECURRENCE

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ABSTRACT - Glossopharyngeal neuralgia with syncope as a sign of neck cancer is a very rare condition. A review of the literature revealed only 29 cases formerly reported. We present the first Brazilian case of such association. A 68-year-old man presented with paroxysmal excruciating pain over the right side of the neck, sometimes followed by syncope. Given the suspicion of recurrent tumor from a previously treated neck malignancy, a computed tomography scan was performed and a right parapharyngeal tumor was shown. Pain and syncope were successfully controlled with carbamazepine and the patient underwent palliative radiotherapy.

KEY WORDS: glossopharyngeal neuralgia, syncope, neck cancer, recurrence.

Neuralgia glossofaríngea com síncope como um sinal de recidiva de câncer do pescoço

RESUMO - Neuralgia glossofaríngea com síncope como um sinal de câncer do pescoço é uma condição muito rara. Uma revisão da literatura revelou apenas 29 casos relatados anteriormente. Apresentamos o primeiro caso brasileiro de tal associação. Um homem de 68 anos se apresentou com dores paroxísticas insuportáveis no lado direito do pescoço, algumas vezes seguidas de síncope. Dada a suspeita de recidiva tumoral derivada de uma malignidade cervical tratada previamente, realizou-se um exame de tomografia computadorizada que evidenciou um tumor parafaríngeo direito. As dores e as síncopes foram controladas satisfatoriamente com carbamazepina e o paciente foi submetido à radioterapia paliativa.

PALAVRAS-CHAVE: neuralgia glossofaríngea, síncope, câncer do pescoço, recidiva.

According to the second edition of the International Classification of Headache Disorders¹, glossopharyngeal neuralgia is a brief severe stabbing pain felt in the ear, base of the tongue, tonsillar fossa or beneath the angle of the jaw. Thus, the pain is located in the distributions of the auricular and pharyngeal branches of the vagus nerve plus of the ipsilateral glossopharyngeal nerve. It is commonly incited by swallowing, talking or coughing and may remit and relapse in the manner of trigeminal neuralgia.

Glossopharyngeal neuralgia was estimated to be 75 times less frequent than trigeminal neuralgia², and its association with syncope was estimated to occur in 1,84% of the cases³. Although generally cryptogenic, this condition sometimes have a demonstrable cause, including neoplasms, infections, inflammations, trauma, elongated styloid process and vascular compression². Glossopharyngeal neuralgia with syncope as a sign of neck tumor is a very rare condition. A fairly diligent review of the literature revealed only 29 cases formerly reported (Table). We present the first Brazilian case of such association.

CASE

A 68-year-old white man with a history of paralaryngeal and pyriform sinus lymphoepithelioma was treated with radical surgery and radiotherapy about 2 years ago. His tumor was stage T₃ N_{2b} M₀ and he underwent bilateral neck dissection, right hemithyroidectomy, pharyngolaryngectomy with later reconstruction by means of a free microvascular right thigh flap, and tracheostomy. After surgery, he received radiotherapy with a total 40 Gy to the entire neck.

After a two years follow-up without clinical signs of recurrence or metastasis, the patient presented with paroxysmal excruciating pain over the right side of the neck, sometimes followed by syncope for two weeks. The neck pain was described as a sudden knife attack deep under the right angle of the jaw and over the right posterior area of his mouth, usually provoked by swallowing or protrusion of the tongue, which lasted about 40 seconds. These pain attacks recurred several times along the day, sometimes followed by diaphoresis, cold extremities, pallor, rotatory vertigo, culminating in fainting with fast consciousness recovery. The patient used to become drowsy after the syncopes for a few minutes. At physical examination, no recurrence of the tumor in the neck and pharynx was apparent to the head and neck surgeon, in addition the patient did not have a histo-

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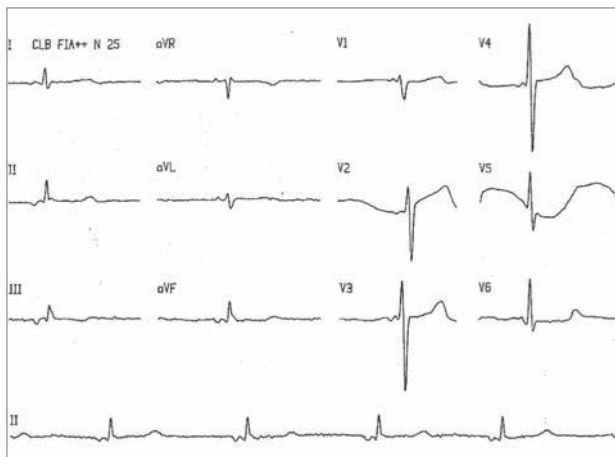


Fig 1. Electrocardiogram showing a temporary bradycardic junctional rhythm after an episode of glossopharyngeal neuralgia with syncope.

ry of syncope or any other cardiovascular disease. However, a typical episode of glossopharyngeal neuralgia with syncope occurred soon after the oral cavity examination had been finished. The patient was immediately referred to the emergency room where he arrived conscious, with 18 respiratory incursions per minute, oxygen digital saturation of 98% on open air, blood pressure of 110/80 mmHg and pulse rate of 50 per minute. Successive electrocardiograms were taken and they showed an initial junctional rhythm, which evolved to a sinusual rhythm without any medical intervention, devoid of any additional symptom or signs of ischemic heart disease (Fig 1).

Witnesses described a tonic posture of the upper limbs prior to the syncope and a subsequent first episode of fecal incontinence, hence the emergency room physician asked for a complete neurological assessment to exclude seizure.



Fig 2. Unilateral atrophy, fasciculations and tongue deviation to the right that were considered to be secondary to right hypoglossal nerve surgical lesion.

Table. Number of reported cases of glossopharyngeal neuralgia with syncope secondary to neck cancer per author(s).

Author(s)	Number of reported cases
Kollmannsberger et al. ¹⁵ (1964)	2
Meienberg et al. ¹⁶ (1975)	1
Roa and Krupin. ¹⁷ (1981)	1
Dykman et al. ¹² (1981)	1
Greenberg et al. ¹⁸ (1981)	1
Sobol et al. ¹⁹ (1982)	1
Fleischmann et al. ²⁰ (1983)	1
Macdonald et al. ⁸ (1983)	2
Kim et al. ²¹ (1985)	1
Weinstein et al. ²² (1986)	1
Chalmers and Olson. ²³ (1989)	2
Papay et al. ⁹ (1989)	3
Pujadas et al. ¹¹ (1990)	1
Metheetrairut and Brown et al. ¹⁴ (1993)	1
Tang et al. ²⁴ (1993)	1
Rumoroso et al. ²⁵ (1996)	1
Carrat et al. ²⁶ (1996)	2
Nakahira et al. ⁷ (2002)	2
Nicholls et al. ²⁷ (2003)	1
Enguita et al. ²⁸ (2003)	1
García et al. ²⁹ (2003)	1
Worth et al. ³⁰ (2005)	1
Ribeiro et al. (current)	1

Apart from expected post surgical and radiotherapeutic alterations, such as mild weakness of the two trapezius muscles and cervical actinic fibrosis, the remainder neurologic examination only showed a unilateral atrophy, fasciculations and tongue deviation to the right (Fig 2). After protruding the tongue to record its alterations, the patient suffered another glossopharyngeal neuralgia without syn-

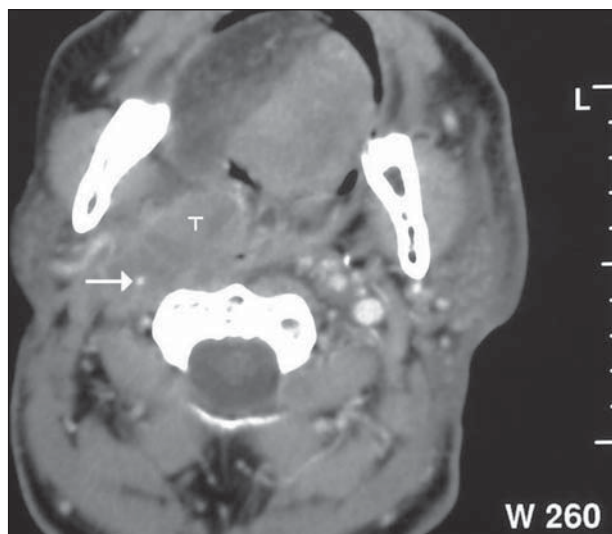


Fig 3. Contrast-enhanced axial computed tomography scan showing right parapharyngeal tumor (T) encasing the right internal carotid artery (arrow) and probably the right

cope, assuming a tonic posture of the upper limbs due to the intense pain. Despite the unawareness of the patient and family of such pathological changes, the right hypoglossal nerve lesion was supposed to be an accidental consequence of the radical neck surgery. Given the suspicion of recurrent tumor from the previously treated neck malignancy, contrast-enhanced computed tomography scans of the head and neck regions were performed and a right parapharyngeal tumor encasing the right internal carotid artery was shown. Besides, a low-attenuating area on the right half of the tongue that represented denervated muscle atrophy and increased fat after the right hypoglossal nerve lesion was also shown, without intracranial lesions (Fig 3). The diagnosis of symptomatic glossopharyngeal neuralgia with syncope as a sign of neck cancer recurrence was made, treatment with carbamazepine initiated and the patient was referred to head and neck cancer reappraisal.

After a few days of treatment, pain and syncope were successfully controlled with carbamazepine 600mg daily, enabling the introduction of a nasogastric tube for nutritional purposes. Carotid sinus massage did not notably cause a decrease in either blood pressure or pulse rate, moreover maneuvers that seemed to trigger the symptoms in the past no longer worked. At the time of writing, the patient had been relieved of glossopharyngeal neuralgia with syncope for about two months and was undergoing palliative radiotherapy.

The patient allowed this report by a written informed consent.

DISCUSSION

As aforementioned, glossopharyngeal neuralgia alone is a relatively uncommon condition², its association with syncope is considered to be very rare³, and this paper reports the thirtieth worldwide case of glossopharyngeal neuralgia with syncope secondary to neck cancer and the first Brazilian case of such association. Among the Brazilian cases of classical glossopharyngeal neuralgia previously reported, four were associated with syncope⁴⁻⁶. Likewise, syncope from head and neck cancer is also considered an exceptional event, ranging from 0,33%⁷ to 0,4%⁸, however, among those cases, glossopharyngeal neuralgia is a frequent cause of syncope due to head and neck cancer invasion, varying from 11,75%⁸, 50%⁷, to 60%⁹. These relatively high frequencies emphasize the need of special imaging techniques to uncover occult malignancies of retropharyngeal and parapharyngeal spaces affecting the glossopharyngeal nerve.

Although limited to few reports, glossopharyngeal neuralgia with syncope due to head and neck cancer becomes a matter of urgency in treatment, because of the cardiovascular complications and advanced cancer stage. Two cardiovascular manifestations occur in glossopharyngeal neuralgia caused by the strong

afferent stimulation activating brainstem mechanisms involved in cardiovascular regulation^{10,11}. One efferent response is the bradycardia or even asystole demonstrated in some reported cases, probably due to vagal discharge, since it is blocked by atropine. Local infiltrations demonstrated that not only the vagus ipsilateral to the pain is activated, but rather a bilateral response occurs, leading to both atrial standstill and prolonged delayed ventricular escape¹⁰.

Separately, the afferent glossopharyngeal stimulus causes inhibition of vasomotor centers, which lead to peripheral vasodilatation and pronounced blood pressure falls, even when the heart is externally paced. Assuming there is no decline in myocardial contractility, cardiac output is diminished despite pacing, probably mainly because of peripheral pooling and decreased venous return¹⁰. Inhibition of sympathetic vasoconstrictor tone may produce those alterations, consistent with the findings of cessation of electrical activity of sympathetic nerves and suppression of neural secretion of norepinephrine during painful episodes leading to hypotension^{12,13}.

Glossopharyngeal neuralgia secondary to recurrent malignant tumor in the neck is considered to carry a poor prognosis when compared to other causes. Intracranial division of the nerve has been advocated as the operation of choice for complete and permanent reduction of pain and cardiovascular symptoms, but this also carries considerable risk, especially in elderly patients. However, medical therapy, including high-dose carbamazepine, should be tried first, particularly in individuals who represent a high operative risk^{13,14}.

We report a case of glossopharyngeal neuralgia with syncope that was associated with occult recurrence of neck cancer in the parapharyngeal space. The mechanism may be linked to invasion of the glossopharyngeal nerve adjacent to the internal carotid artery. In our case, carbamazepine was able to control the symptoms. Careful and repeated investigation of the parapharyngeal space should be required when a patient with previous neck malignancy presents with glossopharyngeal neuralgia with syncope.

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