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Clinical Record

Jugular foramen schwannoma presenting with glossopharyngeal neuralgia syncope syndrome

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Abstract

Introduction: Jugular foramen schwannomas are rare skull base tumours which typically have a variable clinical presentation. Glossopharyngeal syncope syndrome is an unusual clinical presentation; in the following case report, it was the sole presentation of an extracranial jugular foramen tumour.

Methods: The presentation of a patient with glossopharyngeal neuralgia syncope syndrome is reviewed and the pathophysiology, clinical features and treatment discussed.

Results: A 45-year-old woman presented with unilateral throat pain, bradycardia and hypotension leading to episodes of impaired consciousness when lying on her left side or turning her head to the left. Imaging detected a left-sided extracranial jugular foramen schwannoma. The tumour was excised, and the patient had no more syncopal attacks.

Conclusion: Glossopharyngeal neuralgia syncope syndrome can be the sole presentation of a jugular foramen schwannoma. Although this syndrome may be treated with anti-dysrhythmic drugs, cardiac pacing or nerve section, in the presented patient excision of the jugular foramen schwannoma was successful in preventing further episodes of syncope.

Key words: Glossopharyngeal Nerve Diseases; Cranial Nerve Neoplasms; Syncope; Neurilemmoma; Skull Base Neoplasms

Introduction

Jugular foramen schwannomas are rare. They represented only 2.9 per cent of all intracranial neuromas encountered at the National Hospital for Neurology and Neurosurgery over a 20-year period, in a recent review.[1] While the nerve of origin can at times be obscure, vagal schwannomas are reported to constitute half such cases, with glossopharyngeal tumours being infrequent.[2] Imaging findings have led to tumours being classified according to their extracranial, jugular foramen and intracranial components.[3] While intracranial tumours may be more likely to be associated with deafness, ataxia and vertigo, extracranial tumours can present with lower cranial nerve palsies,[4] although the presentation can be varied and indistinct.[1]

Glossopharyngeal neuralgia with syncope is an uncommon syndrome and may be due to a neck mass (often malignant),[5–8] vascular compression of the IXth cranial nerve,[9,10] vascular disease[11] or a foreign body.[12] In the majority of cases, the cause is never found.[13] In a Mayo Clinic series of 217 cases of glossopharyngeal neuralgia, four presented with syncope.[14] The reported incidence is 0.8/100 000.[15]

We report below an unusual case of a patient presenting with glossopharyngeal neuralgia and syncope as the result of a jugular foramen schwannoma.

Case report

A 45-year-old woman presented to the National Hospital for Neurology and Neurosurgery, London, with syncopal attacks of increasing frequency. In the previous two months, she reported suffering six episodes beginning with a sharp pain affecting her left ear, throat and neck. When sleeping on her left side, she would awake with pain feeling nauseous and dizzy. Her light-headedness worsened with head movements to the left. She would then faint, with impaired consciousness for 5–10 minutes. There was no history of seizures. There were no problems with the patient's voice or swallowing, and she did not complain of tinnitus.

On examination, there was very slight medial displacement of the patient's left tonsil. No mass was palpable in her neck, and there were no cranial nerve palsies.

An electrocardiogram revealed heart block. Further autonomic testing found that, with prolonged head-up tilt, the patient had a fall in blood pressure and heart rate. Orthostatic hypotension and postural tachycardia syndrome were excluded. Carotid massage, stimulation over various parts of the pharynx, and swallowing all failed to elicit changes in blood pressure or heart rate.

Magnetic resonance imaging (MRI) revealed a $2.7 \times 2.7 \times 4.5$ cm mass within the carotid space which extended superiorly to the pars nervosa of the jugular foramen and which had the imaging characteristics of a schwannoma. The internal jugular vein was partially effaced (Figure 1).

The schwannoma was resected via an extended cervical incision, following precautionary insertion of a temporary cardiac pacing wire lest there be any peri-operative arrhythmias. At surgery, it was immediately apparent that the

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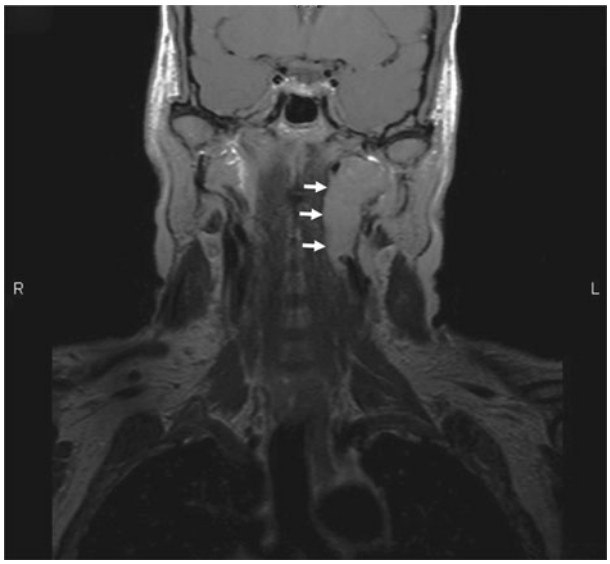

FIG. 1

T1-weighted coronal magnetic resonance imaging scan showing schwannoma (arrowed) within the parapharyngeal space and abutting the jugular foramen. R = right; L = left

vagus was not the origin of the tumour, as it was displaced laterally over its surface (Figure 2). The tumour extended into the jugular foramen and was removed completely in a piecemeal fashion so that collateral damage to adjacent neural structures was minimised. The nerve of origin could not be positively determined, but identification was made via a process of exclusion, by identifying the other carotid sheath structures. The schwannoma seemed most likely to be arising from the glossopharyngeal nerve. Histopathological examination confirmed the diagnosis of a World Health Organization grade one schwannoma.

In the post-operative period, the resection was complicated by a partial and temporary ptosis, and the patient experienced mild dysphagia which subsequently improved significantly. She also had impaired sensation in the distribution of the glossopharyngeal nerve within the oropharynx, which persisted.

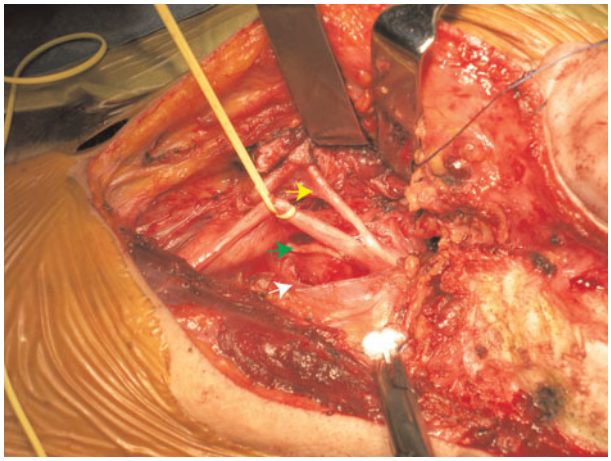

FIG. 2

Surgical photograph showing the schwannoma (bottom arrow) lying medial to the internal jugular vein, which is held in a vascular sling. The hypoglossal nerve (upper arrow) is crossing the bifurcation of the carotid arteries, and the vagus (middle arrow) is seen running over the capsule of the schwannoma.

At the time of writing, the patient had not suffered any further syncopal attacks, and a post-operative MRI scan had confirmed complete removal of the tumour.

Discussion

Jugular foramen schwannomas arise in the medial compartment of the foramen, the pars nervosa, which contains the IXth, Xth and XIth cranial nerves.[16] Symptoms and signs include decreased pharyngeal sensation, impaired gag reflex, palatal deviation, vocal fold palsy with abnormal phonation, sternocleidomastoid and trapezius weakness, deafness, tongue atrophy, hemifacial spasm, ataxia, and hydrocephalus.[1] Imaging confirms a lesion related to the jugular foramen but cannot determine the cranial nerve of origin. A classification scheme has been proposed to aid surgical planning and management.[3]

In 1910, Weisenburg[17] described the occurrence of glossopharyngeal pain in a patient diagnosed at post mortem with a cerebellopontine angle sarcoma. The term glossopharyngeal neuralgia was used in 1921 by Harris,[18] who noted in his paper that Sicard of France had described three similar cases which were treated by nerve section in the neck. In 1942, Riley[19] was the first to report two cases with glossopharyngeal neuralgia, cardiac arrest, syncope and seizures; he described the glossopharyngeal–medullary–vagal 'reflex arc' as the source of the syndrome, and stated it could be blocked by 'cocainization of the throat' and 'atropinization of the vagus'. Taylor *et al.*[13] and St John[20] reviewed a total of 35 cases over the next 40 years.

Chalmers and Olson[5] described 12 cases of glossopharyngeal neuralgia syncope syndrome (also called vagoglossopharyngeal neuralgia) with neck masses; these included five from St John's review, five new cases[8,21,22] and two cases of their own. The neck masses in question comprised nine malignancies, two abscesses and one haematoma. Although a rare clinical syndrome, the importance of glossopharyngeal neuralgia syncope syndrome is exemplified by further reported cases of neck malignancy with syncope and glossopharyngeal neuralgia as the presenting features.[6,7] Similarly, in our patient glossopharyngeal neuralgia syncope syndrome was the sole presentation of a jugular foramen schwannoma.

The key features of glossopharyngeal neuralgia syncope syndrome are unilateral throat, neck or ear pain triggered by swallowing, chewing, coughing, yawning or head movement, with ensuing syncope and seizures, hypotension, and asystole or bradycardia.[11] Cardiac arrest has been reported to last up to 35–40 seconds.[23] Parotid hypersecretion has also been described.[24] In some cases, local anaesthetic applied to the sensitive area in the pharynx can prevent the attack and confirm the diagnosis. Patients with the syndrome have also described a 'tickling sensation' in the throat prior to syncope, as opposed to neuralgia.[25]

In our patient, symptoms were triggered by lying on the left side and turning to the left. A positional trigger has also been described in other cases.[21,23]

Cicogna *et al.*[26] have described a similar presentation in a series of 11 patients with a parapharyngeal lesion; however, glossopharyngeal neuralgia was not a feature and there was no obvious trigger. They termed this phenomenon parapharyngeal space lesion syncope syndrome.

Carotid sinus syndrome with bradycardia and hypotension also presents without pain. Hypersensitivity of the carotid sinus is thought to be the cause, and pressure over the carotid sinus the trigger.[27] Episodes in patients with glossopharyngeal neuralgia syncope syndrome have also been triggered by pressure over the carotid sinus.[11,28]

Central to these three syndromes is an understanding of the carotid sinus reflex, which buffers excessive fluctuations

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in blood pressure. The sinus nerve of Hering arises from baroreceptors in the carotid sinus and joins the glossopharyngeal nerve conveying impulses to the nucleus of the solitary tract in the medulla. If the sinus pressure is raised, thereby increasing baroreceptor stretch tone, inhibition of medullary vasomotor centres results in peripheral vasodilatation, bradycardia, reduced arterial pressure and changes in cerebral circulation.

Signalling from the nucleus of the solitary tract to the dorsal nucleus of the vagus stimulates a parasympathetic cardio-inhibitory response, causing bradycardia via the vagus nerve while independently suppression of the sympathetic outflow, results in a vasodepressor response with peripheral vasodilation and hypotension.[27] A decrease in sympathetic drive has been confirmed by measurements showing decreased secretion of plasma catecholamines[8,23] as well as decreased electrical activity in sympathetic nerves during attacks.[29]

Fainting and loss of consciousness, as seen in our patient, may be explained by cerebral hypoxia following the bradycardia, asystole or hypotension. However, loss of consciousness and seizures have been described in patients who do not demonstrate a change in heart rate and who remain normotensive. This possibly implies that glossopharyngeal stimulation may have an independent effect on cerebral circulation and oxygenation.[27,30]

- **Jugular foramen schwannomas are rare tumours, and glossopharyngeal neuralgia syncope syndrome is an unusual presentation**
- **Although tumours of the neck have been implicated in glossopharyngeal neuralgia syncope syndrome, the majority of these have been malignant**
- **In most cases, this syndrome is treated with a combination of pharmacology and pacing, or with glossopharyngeal nerve section; however, the presented patient was left symptom-free with minimal collateral deficits following removal of the jugular foramen schwannoma**

In glossopharyngeal neuralgia syncope syndrome, it is postulated that pain sensation from the oropharynx, also conveyed by the glossopharyngeal nerve, stimulates fibres carrying sensation from the carotid sinus, either via an ephaptic connection peripherally, or via a central connection in the medulla between the nucleus of the solitary tract and the vagal dorsal motor nucleus.[24,28,29] Ephaptic transmission has been demonstrated experimentally in nerves that are injured or compressed.[31] Electron microscopic findings have also shown degeneration of the myelin sheath in the glossopharyngeal nerves of patients with glossopharyngeal neuralgia.[32] Alpert *et al.*[11] assumed that, in their patient, ischaemia of the glossopharyngeal nerve in the region of the jugular foramen led to the formation of an abnormal motor–sensory connection, with the trigger being swallowing. Compression of the IXth and Xth cranial nerves at the root entry zone can be caused by vascular loops.[10] Cicogna *et al.*[26] postulated that parapharyngeal space lesions caused syncope via abnormal pressure and stimulation of the glossopharyngeal nerve. Sobol *et al.*[21] argued that, in their case, an inflammatory process associated with a malignant tumour and parapharyngeal abscess caused irritation of the glossopharyngeal nerve.

In some cases of glossopharyngeal neuralgia syncope syndrome, atropine can abolish the bradycardia but the hypotension mediated by antisympathetic vasodepressor activity may still persist.[8,23] For this reason, cardiac pacing may also fail.[8,29] Carbamazepine has been used as the sole treatment to control the pain and syncope,[5,12] but has been known to fail after initial success.[13,20] This drug has also been used in combination with cardiac pacing,[7,11,23,28,33] but not always successfully.[8,34] Other drugs have also been tried, such as diphenylhydantoin,[35] belladonna extract,[24] duloxetine[36] and pregabalin,[37] without much success.

Section of the glossopharyngeal nerve, with or without the upper two rootlets of the vagus, generally produces good results in patients with glossopharyngeal neuralgia syncope syndrome[8,20,24,34,35] and parapharyngeal space lesion syncope syndrome.[26] Successful outcomes have also been reported following microvascular decompression.[9,10]

In patients with benign neck masses, such as our reported case, successful outcomes may involve removal of the mass.[21] Temporary cardiac pacing has been used while awaiting surgery, and may be necessary to manage perioperative arrhythmia,[10,34] as in our case.

Conclusion

The case presented is unusual in that jugular foramen schwannomas are rare tumours and glossopharyngeal neuralgia syncope syndrome is an unusual presentation. Although tumours of the neck have been implicated in glossopharyngeal neuralgia syncope syndrome, the majority of these have been malignant. In most cases, this syndrome is treated with a combination of pharmacology and pacing, or with glossopharyngeal nerve section. However, the presented patient was left symptom-free and with minimal collateral deficits following removal of her jugular foramen schwannoma.

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