

Case Report

Glossopharyngeal Neuralgia Caused by Arachnoid Cyst in the Cerebellopontine Angle

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Glossopharyngeal neuralgia is a relatively rare condition characterized by severe, paroxysmal episodes of lancinating pain in the tongue, throat, ear, and tonsil. This disorder is assumed to be due to compression of the glossopharyngeal nerve by vascular structures. A 47-year-old woman complaining of sharp and lancinating pain in the right periauricular and submandibular areas visited our hospital. Swallowing, chewing, and lying on her right side triggered the pain. Her neurologic examination revealed no specific abnormalities. The results of routine hematologic and blood chemistry studies were all within normal limits. Carbamazepine and gabapentin were given, but her symptoms persisted. Her pain was temporarily relieved only by narcotic pain medication. MRI showed an arachnoid cyst located in the right cerebellomedullary cistern extending to the cerebellopontine cistern. Cyst removal was performed via a right retrosigmoid approach. Lateral suboccipital craniotomy was performed using the right park-bench position. After opening the dura and cerebellopontine angle, the arachnoid cyst was exposed. The arachnoid cyst was compressing the flattened lower cranial nerves at the right jugular fossa. Her symptoms resolved postoperatively. Two months after the operation, she was completely free from her previous symptoms.

Key Words : Arachnoid cyst · Cerebellopontine angle · Glossopharyngeal neuralgia.

INTRODUCTION

Arachnoid cysts account for about 1% of intracranial mass lesions^{1,9)}. Arachnoid cysts occur with greater frequency in children than in adults and tend to be discovered more frequently in the left middle cranial fossa^{9,10)}. When the arachnoid cysts are found at the cerebellopontine angle (CPA), they are characteristically located below the cranial nerve (CN) VII and VIII in the posteroinferior aspect¹⁾. Glossopharyngeal neuralgia (GPN) is an uncommon disorder characterized by severe lancinating pain commonly induced by swallowing⁶⁾. The pain is intense and paroxysmal; it originates in the throat, approximately in the tonsillar fossa, and is provoked most commonly by swallowing but also by talking, chewing, yawning, and laughing. The pain may be localized in the ear or radiates from the throat to the ear, implicating the auricular branch of the vagus nerve¹⁸⁾. Most GPN is assumed to be due to the compression of a cranial nerve by the vessels. However, GPNs can also be caused by compression of a nerve by a mass lesion. Several cases of tumors associated GPN have been reported¹⁷⁾ but to date, there has been no report

of GPN caused by an arachnoid cyst. We present the case of a successfully treated 47-year-old woman with GPN caused by an arachnoid cyst.

CASE REPORT

A 47-year-old woman was admitted to our hospital, with a chief complaint of severe pain radiating from the pharynx to the right retroauricular region and to the area beneath the angle of the jaw that persisted last 14 months. Swallowing, chewing, and lying on her right side triggered the pain. Her neurologic examination was entirely normal. Past medical and family histories were unremarkable. Routine hematologic and blood chemistry studies were also normal. Her pain was temporarily relieved only by narcotic pain medication.

Temporal MRI revealed a space-occupying lesion at the right pontomedullary junction, which showed low signal intensity on T1WI, high signal intensity on T2WI, low signal intensity on diffusion-weighted imaging, and no enhancement. It was highly suggestive of an arachnoid cyst located in the right cerebellomedullary cistern extending to the ipsilateral cerebellopontine angle (Fig. 1). Medical treatment with carbamazepine and gabapentin was not effective.

The patient underwent craniotomy for removal of the cyst. Cyst removal was performed via a right retrosigmoid approach. After opening the dura, a large, opaque, thick, and cystic struc-

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ture was noted at the CPA. The dorsal wall of the cyst was excised. The ventral wall of the cyst adhered to and compressed the flattened CNs (VII, VIII, IX, X). The cyst was removed with great care to avoid traumatizing any of the exposed cranial nerves. No vessel compressing the nerves was found in the operative field (Fig. 2).

Her symptoms resolved postoperatively, and the CT scan obtained the day after the operation showed a significantly decreased mass effect in the right CPA. She made an uneventful recovery and was discharged. Two months after the operation, she was completely symptom-free.

DISCUSSION

Arachnoid cysts of the posterior fossa are much less common than those above the tentorium. About 5% of arachnoid cysts are known to be located in the CPA¹⁰. There is a female predominance among patients with cysts in the CPA, with a significant tendency to appear on the right side¹⁰. The mechanism of enlargement of the arachnoid cyst is explained by the osmotic pressure gradient between the inside and outside of the cyst wall, the secretion of fluid by the cyst wall, and possibly by a ball-valve mechanism or inadequate communication of the cystic contents with the subarachnoid space. Enlarging arachnoid cyst produces symptoms by direct compression of the surrounding brain and cranial nerves^{3,7}. Patients with arachnoid cysts may present with ataxia, gait disturbance, headache, nausea, vomiting, character change, memory disturbance, symptoms mimicking transient ischemic attack, dysesthesias, dysphagia, nystagmus, and oscillopsia^{1,12,19}. These cysts also can cause dysfunction of specific CNs, including V (trigeminal neuralgia), VI (diplopia), VII (hemifacial spasm and facial nerve palsy), and VIII (hypacusia, tinnitus, and vertigo)^{3,9,11,12,16}. However, to date there has been no report of a patient with glossopharyngeal neuralgia caused by an arachnoid cyst. Suboccipital craniotomy and cyst excision is recommended for the treatment

of cerebellopontine angle arachnoid cysts. Jallo and colleagues resected the lateral and posterior cyst walls widely and fenestrated the anterior membrane, using microsurgical techniques, into the cisterns surrounding the adjacent cranial nerve¹¹.

The glossopharyngeal nerve is named for its role in providing sensation to the posterior tongue and pharynx; however, it has additional functions. It exits the brainstem as several rootlets along the upper ventrolateral medulla, just below the cranial nerve VIII, between the inferior olive and the inferior cerebellar peduncle. The nerve traverses the subarachnoid space to exit the cranial base via the jugular foramen. It has the following four functional categories; branchial motor, parasympathetic, general somatic sensory and general visceral sensory functions. Its general somatic sensory function is the sensation of touch, pain, and temperature from the posterior one-third of the tongue, in addition to the pharynx, middle ear, and the region near the external auditory meatus².

Idiopathic GPN is a relatively rare condition characterized by severe, paroxysmal episodes of lancinating pain. The pain is similar to trigeminal neuralgia (TN) but is instead localized to the external ear canal, the base of the tongue, the tonsil, or the area beneath the angle of the jaw¹⁸. The first association between GN and vascular compression of the glossopharyngeal nerve was reported in 1889 by Pope¹⁸, who described a patient with pain and loss of taste as a result of compression of the glossopharyngeal nerve by a dilated and thrombosed vertebral artery. Shortly thereafter, in 1936, Lillie and Craig¹⁸ described an

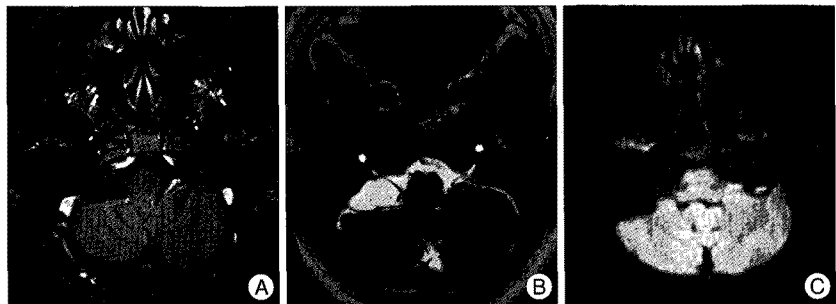


Fig. 1. Temporal MRI reveals a space-occupying lesion in the right cerebellopontine angle, which shows low signal intensity without enhancement at T1-weighted image (A), high signal intensity at T2-weighted image (B), low signal intensity at diffusion weighted image (C). It was diagnosed as an arachnoid cyst located in right cerebellomedullary cistern with extension to the ipsilateral cerebellopontine angle.

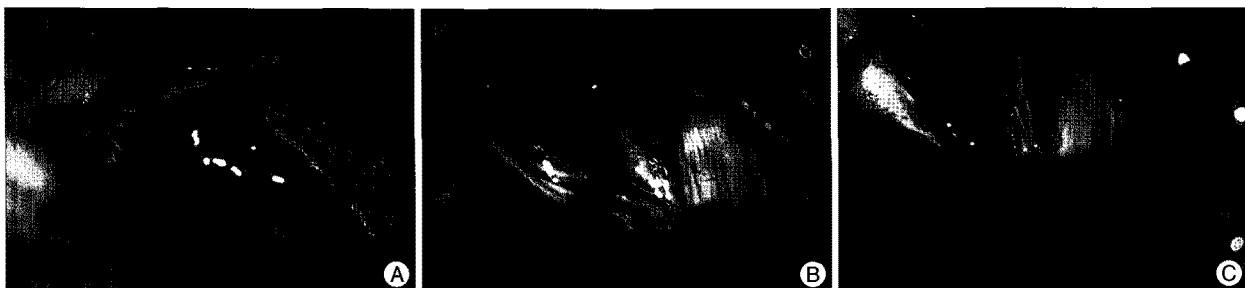


Fig. 2. Intraoperatively, a large, opaque, thick and cystic structure is noted in the cerebellopontine angle (A). The dorsal wall of the cyst is excised and the ventral wall of the cyst appears to be adhered to CNs (VII, VIII, IX, X) and flattens the CNs around right jugular fossa (B). The cyst is excised and decompressed with great care to avoid traumatizing any of the exposed cranial nerves. After removal of the cyst, there is no compressing vessel (C).

anomalous arterial loop as the cause of GPN. Although vascular compression is known as the most common cause, multiple sclerosis, posterior fossa neoplasm, cyst, and trauma may also contribute to GPN. Several pathogenic mechanisms may produce GPN. They include vascular compression, tumors in the CPA, extracranial lesions, and unknown etiology^{15,17}. GPN caused by a CPA tumor has been previously reported; however, it is very rare^{13,17}. In the report on combined hyperactive dysfunction syndrome of the cranial nerves, Kobata et al.¹⁴ reported three cases of epidermoid tumors located in the CPA as a causative factor of the syndrome, but GPN was not present in those cases. To our knowledge, only one case of GPN with the epidermoid tumor in the posterior fossa was reported¹⁷. This is the first case of GPN completely resolved after removal of an arachnoid cyst.

GPN is usually diagnosed by clinical features but neuroimaging study plays a key role in detecting the underlying lesions, such as vascular compression, tumors or demyelinating plaques. MRI can be an extremely accurate tool for detecting neurovascular compression⁴. The combination of combined constructive interference in steady-state imaging and diffusion-weighted imaging proved to be useful in an irritative syndrome involving a cranial nerve that was raised by an intracranial tumor¹⁷.

Several surgical approaches to medically intractable GPN have been described, but most of them based on the destruction of the glossopharyngeal and vagus nerve fibers⁵. More recently, microvascular decompression has been shown to be an effective treatment and should be considered as the first treatment in cases of drug-resistant GPN with vascular compression. Despite new anti-epileptic drugs that have been introduced in the treatment of glossopharyngeal neuralgia, surgery is still needed in the patients who show drug intolerance, refractoriness, or both^{5,8,20}.

In addition to vascular compression, tumor-like lesions may cause hyperactive dysfunction syndromes due to their compression on the cranial nerves. The mass may compress the cranial nerve directly, indirectly through a vascular loop, or by both means^{13,17}. In this case, there were no vessels compressing the glossopharyngeal nerve intra-operatively. The main causes of GPN in this case were direct compression by the cystic mass and adhesion of the cystic wall to the lower CNs.

CONCLUSION

GPN is a rare disorder that commonly occurs most commonly due to vascular compression. The incidence of arachnoid cysts in the CPA is very low, and they usually cause dysfunction of specific CNs, including V, VII, and VIII. However, in this case, GPN developed secondary to an arachnoid cyst in the CPA and cyst removal resulted in complete pain relief.

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