Ganglioneuroma of Glossopharyngeal Nerve in a Patient with Glossopharyngeal Neuralgia: A Case Report

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Ganglioneuroma is a rare tumor. Such tumor arising from cranial nerve is further rare. So far our knowledge, in the literature there is no report of ganglioneuroma involving glossopharyngeal nerve. Here, we report a case of very small glossopharyngeal nerve ganglioneuroma and the patient also had longstanding glossopharyngeal neuralgia (GPN). Case Report: A 40-year-old male diagnosed case of left GPN for last 7 years presented with gradual unresponsiveness of drug for last 5 years. Due to severity of pain sometime, he wished to do suicide. Magnetic resonance imaging (MRI) of head revealed only suspected loop of vessel in root entry zones of 9th and 10th cranial nerves on left side. The patient underwent explorative posterior fossa craniotomy. Careful dissection of arachnoid over 9th cranial nerve near jugular foramen (JF) revealed thick and red color nerve with nodularity (tumor like). Dissection of arachnoid at nerve root entry zones of 9th and 10th nerves also revealed an aberrant loop of posterior inferior cerebellar artery (PICA). The 9th nerve was transected and suspected “tumorous portion” of nerve was sent for histopathological examination. The PICA loop was dissected away from root entry zones by placing muscle and surgical between 10th nerve roots and PICA loop. He made an uneventful recovery. Histopathological examination confirmed ganglioneuroma. Six months after the operation, he was free of symptoms. In this case, probably previously existing GPN was worsened by the growth of ganglioneuroma and surgical treatment brought gratifying result.

Keywords: ganglioneuroma, glossopharyngeal nerve, intractable, glossopharyngeal neuralgia, aberrant vascular loop

Introduction

Ganglioneuroma is a neural crest origin tumor commonly occurs in mediastinum, adrenal glands, and peripheral nervous system but very rarely it occur in cranial cavity.1–3 Very few cases of intracranial ganglioneuroma arising from the trigeminal nerve have been reported in the literature.2,3,5 Glossopharyngeal neuralgia (GPN) is a rare clinical condition characterized by intense lancinating pain in throat commonly triggered by swallowing6 but also by talking, chewing, yawning, and laughing. The pain is severe and paroxysmal; it originates in the throat, approximately in the tonsilar fossa. It may be localized in the ear or radiates from the throat to the ear, indicating the auricular branch of the 10th cranial nerve.3 Most GPN is assumed to be due to the compression of a cranial nerve by the vessels, commonly by posterior inferior cerebellar artery (PICA) or vertebral artery (VA). However, GPNs can also be caused by compression of a nerve by a mass lesion. There is several reports where tumors are associated GPN1,2,3,6 where ix nerve is compressed by extrinsic large size tumor in cerebello-pontine angle (CPA) but till today, there is no report of glossopharyngeal nerve ganglioneuroma or gangliomeuroma of ix nerve with pre-existing GPN. Here, we report a case of very small glossopharyngeal nerve ganglioneuroma near jugular foramen (JF) and the patient also had longstanding GPN.

Case Report

A 40-year-male presented with recurrent sudden, severe lancinating, short lasting (15–20 seconds) pain in throat for last 11 years and he was a diagnosed as left GPN. The pain was precipitated by taking food (swallowing/chewing) or drink from left side of the throat and then to retro-mandibular region and ear. Sometimes, due to severity of pain, he wished to do suicide. There was history of series of pain event, that is, 30–40 times pain attacks within days to week. Then, it might remain silent for several weeks and again recur irrespective of taking drug. Initially, the pain responded well to carbamazepine especially for last 5 years. His neurological and other systemic examinations were normal. Past medical and family histories were unremarkable. Routine laboratory investigations including X-ray cervical spine (including head–neck junction) were also normal. Magnetic resonance imaging (MRI) of head 3D CISS (FIESTA) images revealed suspected loop of vessel in root entry zone of 9th and 10th cranial nerves on left side (Fig. 1). Left-sided glossopharyngeal nerve was thick and irregular in comparison to right side (Fig. 1C) (detected by reviewing after operation).

Operation

The patient underwent explorative retromastoid, retrosigmoid lateral suboccipital craniotomy in park bench position keeping left side up under general anesthesia. After opening

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the dura, lower CPA was exposed. Lower cranial nerves were
exposed and identified after incision and dissection of arach-
noid over it. There was some arachnoid thickening and
scaring on 9th cranial nerve near upper compartment of JF.
After careful dissection over 9th cranial nerve near JF, the
nerve was found thick, red and nodular (tumor like). Further
dissection of arachnoid at nerve root entry zone of 9th and
10th nerves revealed an aberrant loop of PICA (Figs. 2A and
2B) The 9th nerve was transected and suspected “tumorous
portion” of nerve was sent for histopathological examination
(Figs. 2C–2F). The PICA loop was dissected away from root
entry zones (both 9th and 10th cranial nerves) by placing
muscle-fascia and surgical between 10th nerve roots and
PICA loop.

Postoperative period
His symptoms resolved postoperatively but developed low-
pitched voice with hoarseness without any swallowing
problem that recovered completely within 10 weeks after
operation. The computed tomography (CT) scan obtained on
the 1st postoperative day showed nothing abnormal other
than the post craniotomy state. He made an uneventful
recovery and was discharged. Histopathological examination
revealed ganglioneuroma (Fig. 3). Immunohistochemistry
staining showed S 100, GFPA, NSE, NFP, and Chromog-
rarin positive and Cytokeratin, Vimentin & CD31 negative,
which confirmed ganglioneuroma. Six months after the
operation, he was completely symptom-free.

Discussion
In idiopathic GPN, the pain is similar to trigeminal neu-
ralgia but it is localized to the ear (external), the base of
the tongue, the palatine tonsil, or the area behind the mandibular
angle. In 1889, Pope first reported the association between
glossopharyngeal nerve and vascular compression of the 9th
cranial nerve roots. He described a case with pain and loss of
taste sensation from compression of the 9th cranial nerve by a
dilated and thrombosed VA. Lillie and Craig in 1936
described an anomalous arterial loop as the cause of GPN.
Although aberrant vascular compression is the most common
etiology, multiple sclerosis, posterior fossa tumor, cyst, and
trauma can also cause GPN. Several pathogenic mechanisms
may produce GPN. So aberrant vascular compression, neo-
plasms in the CPA, extracranial lesions, and unknown etiology
may cause GPN. GPN due to a CPA neoplasm (such as epi-
dermoid) has been previously reported; however, it is very rare
and usually occurs due to compression on glossopharyngeal
nerve by a large cerebello-pontine tumor. Our case is the
first case of GPN completely resolved after removal gangli-
oneuroma with glossopharyngeal neurectomy as well as
micro-vascular decompression (MVD) to decompress the
glossopharyngeal nerve roots with vagal nerve roots. Here, we
think ganglioneuroma was just incidental development of

tumor in the patient with pre-existing neuralgia and it might
worsened the neuralgic sufferings of the patient.

Ganglioneuroma is a rare and benign tumor of the auto-
nomic nerve fibers arising from neural crest sympatho-
gonia, which are completely undifferentiated cells of the
sympathetic nervous system. However, ganglioneuromas
themselves are fully differentiated neuronal tumors that do
not contain immature elements. Ganglioneuromas most
frequently occur in the abdomen; however, these tumors
can grow anywhere sympathetic nervous tissue is found.
Other common locations include the adrenal gland, para-
spinal retroperitoneum, posterior mediastinum, head, and
neck.

Intracranial ganglioneuroma is very rare. Only few cases of
ganglioneuroma involving 5th cranial nerve were
reported. Ganglioneuroma involving other cranial nerves
including glossopharyngeal nerve was not reported.

Commonly GPN is a clinical diagnosis but neuroimaging is
used in identification of the underlying pathology, such as
aberrant vascular compression, neoplasms, or demyelinating
plaques. High-resolution MRI is very sensitive tool for
detecting aberrant vascular compression. In this case, MRI
could suspect that an aberrant vascular loop may compressing
the nerve root but failed to identify the ganglioneuroma arising
from 9th cranial nerve roots near JF as found per operatively.
After operation when we reviewed the MRI images, we found
that the nerve roots near JF were irregular and thick.

Fig. 1  (A and B) MRI of head 3D CISS/FIESTA images showing suspected loop of vascular loop at the root entry zones of left glossopharyngeal
and vagal nerve roots (arrow marked); ix: glossopharyngeal nerve. (C) MRI of head 3D CISS/FIESTA images showing thick irregular margins
glossopharyngeal nerve on left side in comparison to right side. White arrow marks showing 9th cranial nerve and orange arrow marks suspected
vessel loop. ix: glossopharyngeal nerve.
Ganglioneuroma of Glossopharyngeal Nerve

Despite new anti-epileptic drugs in the treatment of GPN, microsurgery is needed in the patients who show drug intolerance, refractoriness, or both. Several surgical treatments to medically intractable GPN based on the destruction of the glossopharyngeal and vagus nerve fibers are now rarely used. Nowadays, MVD is an effective treatment and should be used as the first treatment in drug-resistant GPN with aberrant vascular compression.

In addition to aberrant vascular compression, neoplasm-like conditions originating from nerve itself may cause hyperactive dysfunction syndromes by compression and irritation of its own nerve fibers and fascicles. In our case, causes of GPN were due to an aberrant vessel (i.e., PICA) compressing the glossopharyngeal nerve (confirmed intra-operatively) and ganglioneuroma originating from glossopharyngeal nerve roots near JF (found during operation and identified retrogradely on MRI) might have worsened the GPN instead of being the etiology of GPN.
In this case, glossopharyngeal ganglioneuroma developed in pre-existing GPN patient, which might further increase the woe of the neuralgia and surgical treatment brought gratifying result.

Conflicts of Interest Disclosure
There are no conflicts of interest and nothing to disclose.

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