Case Report

Vagoglossopharyngeal Neuralgia Occurred Concomitantly with Ipsilateral Hemifacial Spasm and Versive Seizure-Like Movement: A First Case Report

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Vagoglossopharyngeal neuralgia (VGPN) is a very rare condition. VGPN with convulsive like attack is even rarer. All of the cases had their head turned to the opposite side of facial pain. Hemifacial spasm occurring concurrently with VGPN has never been reported. Herein, we present the first case of VGPN that had ipsilateral hemifacial spasm and versive seizure-like movement to the same side of facial pain. We reported a 71-year-old man presenting with multiple episodes of intermittent sharp shooting pain arising on the right middle neck, followed by hemifacial spasm on right face. Then the patient became syncope while his head and gaze turned to the same side of the painful neck. Electrocardiography showed sinus arrest. Interictal Electroencephalography was normal. This patient initially responded to pregabalin for two weeks, then the symptoms became worse. Microvascular decompression and carbamazepine resulted in the complete remission of all symptoms after six months of follow-up. We could not explain the pathophysiology of unilateral versive seizure like movement.

Keywords: *Vagoglossopharyngeal neuralgia, Cardiac syncope, Versive seizure-like movement, Microvascular decompression*

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Glossopharyngeal neuralgia (GPN) is a rare condition account for 0.2 to 1.3% of facial pain syndrome. Generally, it presents with unilateral paroxysmal severe stabbing or lancinating pain in the throat, ear, tonsillar area, angle of mandible, or neck. Occasionally, GPN is associated with cardiac syncope[1-3], so it can be named "vagoglossopharyngeal neuralgia" (VGPN). Many reviews revealed various electrocardiography (ECG) findings, such as sinus bradycardia, asystole[4], ventricular escape[5], bradyarrhythmia, sinus arrest[6], and normal ECG with hypotension[7]. There are few reports on convulsive-like attacks with syncopal attack[7-9]. However, several patients had their heads turned to the opposite side of facial pain[5,7]. Due to lack of data, the pathophysiology of this versive seizure-like movement could be explained by the same mechanism as hemifacial spasm. Ephaptic transmission and ectopic excitation of the accessory nerve could be the cause of sternocleidomastoid muscle spasm, and this may result in the patient's head turning to the contralateral side of the painful neck. The electroencephalography (EEG) mostly appeared normal. Various treatment options have been reported to be effective, such as carbamazepine, gabapentin, pregabalin, and surgical treatment (microvascular decompression or selective rootlet rhizotomy). Temporary or permanent pacemaker is also useful adjunct treatment for preventing life threatening syncope.

We report a case of VGPN who had a neck pain and transient loss of consciousness occurring with ipsilateral versive seizure-like movement. The patient initially responded to pregabalin for two weeks, then the symptom became worse. We successfully treated him with microvascular decompression and carbamazepine.

Case Report

A 71-year-old man presented with intermittent sharp shooting neck pain and transient loss of consciousness for four months. Each spell started with pain at the middle part of the right side of the neck followed by transient loss of consciousness,

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approximately five to ten seconds. The episode could be provoked by eating or drinking. Furthermore, the attacks occurred from twice weekly to daily, but sometimes the attack happened as frequent as ten times per day. This patient had been diagnosed as GPN. Oral pregabalin was started at 75 mg/day and was effective in controlling the symptoms. Neither spell nor neck pain occurred during the first two weeks. Unfortunately, he developed multiple episodes of intermittent neck pain, 20 seconds each, followed by right hemifacial spasm and nausea. Then, he became unconscious and his head turned to the right, which was similar to versive seizure. His right arm and left leg were spastically flexed. Simultaneously, his left arm and right leg were spastically extended. The spell took only five to ten seconds, then he regained consciousness (Fig. 1).

Physical examination and Neurological examination were normal as were Interictal EEG finding. ECG revealed sinus arrest three to five seconds during syncope. Cranial magnetic resonance imaging showed right posterior inferior cerebellar artery impinged to root of exit of cranial nerves IX, X, and XI (Fig. 2).

We started treatment with carbamazepine and slowly titrated up to the maximum dosage. However, the spells occurred more frequently and the sinus arrest took longer period, so we decided to implant an intravenous temporary pacemaker as an additional treatment. Despite the pacemaker implantation, patient continued to have intermittent transient loss of consciousness. We contemplated right suboccipital craniotomy and microvascular decompression due to the refractory medical treatment. Operative findings revealed the loop of right posterior inferior cerebellar artery (PICA) that impinged cranial nerves IX, X, and XI at the root exit zone (Fig. 3). The glossopharyngeal nerve was the most severely compressed by PICA, and the accessory nerve obtained a pressure effect from vascular pulsation. Tight adhesion between the glossopharyngeal nerve and the accessory nerve was observed. Microsurgical dissection was performed to separate nerves from PICA. Then we placed Teflon between the nerves and the artery as a shock absorber. There were no postoperative complications. On the first postoperative day, the patients still develops three spells. From the second postoperative day, carbamazepine was started and effectively controlled the spells. This patient had no further attacks and all symptoms were resolved after six months of follow-up.

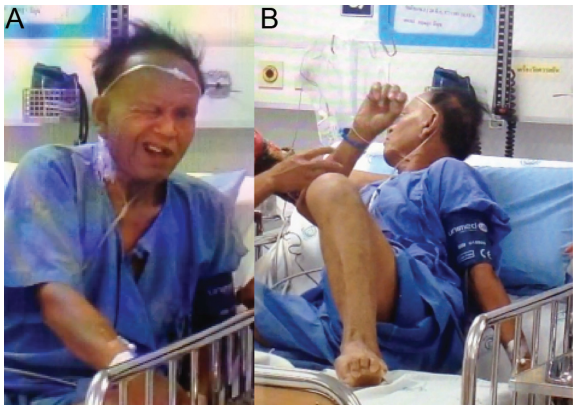

Fig. 1 A) Clinical manifestation during spell showed right hemifacial spasm preceded by sharp shooting pain at the right middle neck. B) Versive like movement.

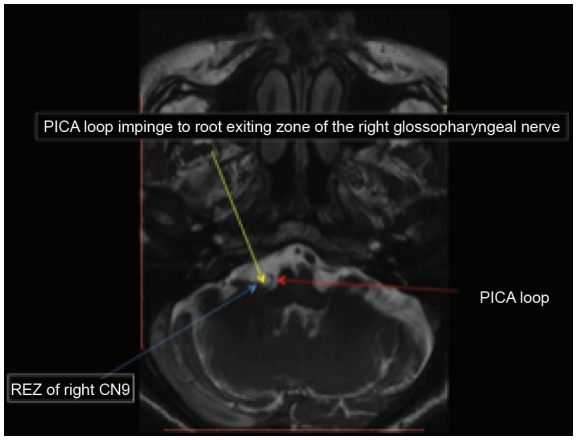

Fig. 2 MRI finding showed posterior inferior cerebellar artery loop impinges to right glossopharyngeal nerve at root exiting zone.

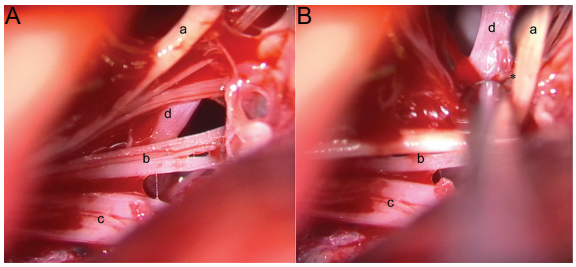

Fig. 3 A) Posterior inferior cerebellar artery loop compressed CN IX, X, and XI from the anterior. Accessory nerve adheres to glossopharyngeal nerve. B) Tight adhesion (*) between accessory nerve (a) and posterior inferior cerebellar artery (d). Operative findings; a = accessory nerve, b = vagus nerve, c = glossopharyngeal nerve, d = posterior inferior cerebellar artery.

Discussion

VGPN is a rare disease account for 2% of GPN. Many studies reported the clinical symptoms of transient loss of consciousness concomitant with pain arising from glossopharyngeal nerve distribution as syncope, cardiac syncope[10-13]. Neither hemifacial spasm nor head movement to the same side of facial pain concurrent with GPN has been reported.

There are also few reports about abnormal convulsive-like movement that occurred during pain attack[4,14-17]. Bruyn[2] reported 304 cases of GPN: 11 cases had cardiac syncope but only one patient had convulsive movement. This versive convulsion-like movement, characterized by turning head and gaze to the opposite side of neck pain, was reviewed in some literatures[5,7]. Most patients were investigated by EEG for excluding seizure. Most EEGs appeared normal including our patient. Varrasi et al[7], reported a progressively high amplitude slowing pattern on the EEG.

Most common ECG finding in VPGN is bradycardia followed by asystole. Johnston and Redding[6] reported VGPN with sinus arrest. Kazemi and Akbarzadeh[5] also reported asystole followed by very slow ventricular escape beat during syncope. This patient's ECG could be explained by the right vagus nerve stimulation causes sinoatrial node stimulation, which resulted in sinus arrest.

Gaul et al[18] reported MRI findings in 19 cases of GPN. All cases had neurovascular compression syndrome of the glossopharyngeal nerve and vagus nerve, but only three cases had syncope.

When compared with the trigeminal neuralgia and hemifacial spasm, VGPN is probably caused by ephaptic transmission and ectopic excitation of glossopharyngeal nerve. The excessive input from glossopharyngeal nerve may be transmited via the nucleus of the solitary tract and then activates the dorsal motor nucleus of the vagus nerve. This probably results in parasympathetic overactivity and inhibits the sympathetic activity. Likewise, activation of facial nerve via the nucleus of the solitary tract could result in hemifacial spasm. On the other hand, the mechanism of the activation of accessory nerve remains unclear. From our intraoperative finding, we hypothesize that the accessory nerve may have abnormal synapse with the glossopharyngeal nerve. An ectopic excitation of the glossopharyngeal nerve transmits via abnormal synapse and causes the accessory nerve excitation, which results in the sternocleidomastoid muscle spasm with turning of the head to the opposite side to neck pain.

The variation of symptoms in GPN with/without syncope or seizure like movement, can be explained by anatomical variation of the PICA loop that compresses these cranial nerves. If there was a compression of both the glossopharyngeal nerve and the accessory nerve, this might increase the propensity to have adhesion between nerves leading to an abnormal synapse.

In the present report, the patient turned his head to the same side of neck pain. We could not explain the mechanism of this symptom due to the supranuclear innervation of the accessory nerve remained not yet clear. However, we tried to formulate three hypotheses to explain the mechanisms of ipsilateral versive-like movement. First hypothesis is the direct excitation of ipsilateral accessory nerve through its connection to the vagus nerve at its upper portion, results in trapezius muscle spasm. When the scapula is fixed, trapezius muscle contraction causes head turns ipsilaterally with the face turns upward as we seen in this patient. The second hypothesis is the excitation nucleus of tractus solitarious transmits through its complex connections to contralateral spinal accessory nucleus, results in contralateral sternocleidomastoid muscle spasm. The variation of the symptoms depends on the anatomical variation of the central myelin portions of the cranial nerves[19]. Third hypothesis, we define this symptom as complex partial seizure. His spell started with versive-like movement then followed by abnormal flexion of his right arm and extension of his left arm as motor sign of seizure. Moreover, he also had cloudy of consciousness simultaneous with the motor signs, meanwhile, ECG showed normal pacemaker rhythm. He still had confusion for few minutes after his head and arms turn to normal posture, that similar to post-ictal state. Consequently, we thought that his symptom could be complex partial seizure. We hypothesized that mechanism of seizure probably cause by global cerebral hypoxia. Ectopic excitation of glossopharyngeal nerve could activate the nucleus of tractus solitarious results in rapidly increase in vagal tone and suddenly hypotension. Transient global cerebral hypoxia may occur, and could trigger the seizures. Perhaps, patient had underlying cause of seizure near his left rolandic area that could not be identified by MRI and interictal EEG.

The GPN can be treated by medication or surgery. The first line of treatment is medication. Carbamazepine is usually chosen as the first line of medication. If the pain is uncontrollable, the second

line drugs, such as pregabalin, gabapentin and duloxitine are also effective[4,20,21].

Temporary transvenous cardiac pacemaker implantation should be considered if patients had frequent syncope[12,21-23]. Although the temporary pacemaker cannot decrease the frequency of syncope, we suggest that it can prevent life threatening syncope. A permanent pacemaker should be considered in patients with hypersensitive carotid sinus syndrome or long-standing, relapsing, severe syncope[6,17,21,23]. We decided to implant the temporary pacemaker due to an increase in the frequency of syncope and for operational safety.

Surgical treatment should be contemplated if the patient was refractory to medical treatment. Chen and Sindou[24] reviewed 515 patients with GPN who had undergone microvascular decompression and demonstrated a total pain relief rate of more than 90% with a lower recurrent rate compared with rhizotomy and stereotactic radiosurgery. However, stereotactic radiosurgery has the advantage of its lower risk of surgical complications, such as dysphagia, hoarseness of voice or facial paresis associated with permanent nerve damage. By the six-month follow-up, this patient had achieved complete remission with the microvascular decompression and carbamazepine.

What is already known on this topic?

Convulsive like movement occurred during pain attack is a rare presentation of vagoglossopharyngeal neuralgia that could be described its pathophysiology by anatomical variation of the posterior inferior cerebellar artery compressing the glossopharyngeal nerve or the vagus nerve, as well as the spinal accessory nerve. Consequently, the nerves that are compressed have ephaptic transmission and ectopic excitation result in symptom of cranial nerve overactivity.

What this study adds?

This report showed an atypical presentation of VGPN that could not be explained its neural pathway. Discovering supranuclear connection between the nucleus of tractus solitarious and the contralateral spinal accessory nerve in the future may be useful for describe atypical symptoms of the brainstem's lesions.

Informed consent

Patient was provided their informed consent for the publication of this case report.

Potential conflicts of interest

None.

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***Vagoglossopharyngeal neuralgia* พบร่วมกับใบหน้ากระตุกและอาการคล้ายชักชนิด *versive* โดยหันศีรษะไปข้างเดียวกับใบหน้าข้างที่เจ็บ: รายงานผู้ป่วย**

พีระพงศ์ เธียราวัฒน์, อภิรัตน์ หวังธีระประเสริฐ, จิราพร จิตประไพกุลศาล

Vagoglossopharyngeal neuralgia (VGPN) เป็นภาวะที่มีอุบัติการณ์น้อยมาก และ VGPN ที่พบร่วมกับอาการที่คล้ายการชักนั้นยิ่งหาได้ยาก โดยอาการชักที่พบร่วมด้วยผู้ป่วยจะหันศีรษะไปด้านตรงกันข้ามกับอาการเจ็บที่ใบหน้า ปัจจุบันยังไม่มีรายงานอาการใบหน้ากระตุกที่พบร่วมกับ VGPN รายงานผู้ป่วยฉบับนี้นำเสนอผู้ป่วย VGPN รายแรกที่มีอาการใบหน้ากระตุกและมีอาการคล้ายชักชนิด versive โดยผู้ป่วยหันศีรษะไปทางเดียวกับข้างที่เจ็บหน้า

คณะผู้นิพนธ์ได้เสนอผู้ป่วยชายไทยอายุ 71 ปี มีอาการเจ็บบริเวณลำคอข้างขวา ลักษณะอาการเจ็บเหมือนถูกเข็มแทงตามด้วยใบหน้าซีกขวากระตุก หลังจากนั้นผู้ป่วยเริ่มไม่รู้สึกตัวพร้อมกับหันศีรษะไปข้างขวาและตากลอกไปทางขวา ซึ่งเป็นข้างเดียวกับใบหน้าข้างที่เจ็บโดยมีอาการเป็น ๆ หาย ๆ ผลการตรวจคลื่นไฟฟ้าหัวใจพบว่า มี sinus arrest ผลการตรวจคลื่นไฟฟ้าสมองพบว่าปกติ ผู้ป่วยตอบสนองดีกับการรักษาขั้นแรกด้วยยา pregabalin แต่ 2 สัปดาห์ต่อมา พบว่าอาการแย่ลง จึงตัดสินใจรักษาด้วยการผ่าตัด microvascular decompression และให้ยา carbamazepine รับประทานต่อเนื่อง 6 เดือน ซึ่งผู้ป่วยไม่มีอาการดังกล่าวอีก ไม่เคยมีรายงานผู้ป่วย vagoglossopharyngeal neuralgia ที่มีใบหน้ากระตุก และ มีอาการคล้ายการชักชนิด versive โดยหันศีรษะไปข้างเดียวกับข้างที่เจ็บหน้า และยังไม่สามารถอธิบายพยาธิสรีรวิทยากำเนิดของอาการเหล่านี้ได้