



## CASE REPORT

**A CASE REPORT ON WEISENBERG SYNDROME DUE TO VASCULAR COMPRESSION****Harikrishnan K<sup>1\*</sup>, Anjaly S<sup>1</sup>, Maria C Kuriakose<sup>1</sup>, Bhaghya Prasad<sup>1</sup>, Bimal Govind<sup>2</sup> and Anil Babu A<sup>1</sup>**<sup>1</sup>Department of Pharmacy Practice, National College of Pharmacy, Manassery, Calicut, Kerala, India.<sup>2</sup>Neurosurgeon, KMCT Medical College Hospital, Manassery, Calicut, Kerala, India**Received for publication:** June 07, 2015; **Revised:** June 10, 2015; **Accepted:** June 18, 2015

**Abstract:** Weisenberg syndrome is an important facial pain syndrome due to its rarity as well as the difficulty in diagnosing the condition. It is often misdiagnosed as trigeminal neuralgia. The pain experienced in Weisenberg syndrome is extremely disturbing for the patients and the condition is potentially fatal due to its association with cardiac syncope, arrhythmias and hypotension. We report a case of Weisenberg syndrome in a male patient presented with one sided throat pain with auricular involvement. We discuss the currently preferred surgical and non-surgical options in the treatment of the condition.

**Key word:** Glossopharyngeal Neuralgia; Microvascular Decompression; Neurectomy; Oxcarbazepine

**INTRODUCTION**

Weisenberg syndrome is a rare pain syndrome affecting the glossopharyngeal nerve, which is usually unilateral in origin and characterized by intense and paroxysmal pain localized to the ear, base of the tongue, back of the throat, beneath the angle of the jaw and it can radiate down the neck. It is also known as Glossopharyngeal Neuralgia (GPN). Vascular compression of the glossopharyngeal nerve root and the upper rootlets of vagus nerve results in glossopharyngeal neuralgia. The excruciating pain may last for a few seconds to a few minutes. Major symptoms are severe pain in one side of throat, syncope, hypotension, palatal myoclonus and bradycardia. The disease can be managed by drugs and surgery.

**CASE REPORT**

A 55-year old male patient with no known comorbidities presented with severe right sided throat pain radiating to ear, in the neurosurgery department of a tertiary care teaching hospital. The repeating episodes of unbearable pain which is increasing in intensity was found to last for about 10-15 seconds. Medical history revealed that the pain appeared about 12 months prior to his admission in the hospital. The pain was exaggerated on swallowing and radiated to the back of the tongue and towards the right side of ear. Even though the pain had a recurring nature, it was absent between attacks. He experienced weakness and fatigue as he was not taking food regularly due to intense pain on swallowing and chewing. For the last 3 months he regularly administered IV dextrose from a nearby health care center.

During the first episode of pain, our patient consulted a physician and was prescribed pain killers for 2 months. But, still the pain persisted and he visited another clinician, who advised him to take rabeprazole

for a period of 2 months, as the symptoms were indicative of Gastroesophageal Reflux Disease (GERD) associated GI disturbances. Later, the patient noted that the pain was increasing in intensity and he consulted an ENT specialist who diagnosed the condition as migraine. He was then prescribed T. Propranolol (migraine prophylaxis) and T. Vasograin (Ergotamine tartrate 1 mg, Caffeine 100 mg, Paracetamol 250 mg, Prochlorperazine maleate 2.5 mg) for a period of 6 months. But, during the course of therapy patient informed the physician about the recurring nature of pain and thus he was referred to a neurologist. The patient consulted the neurologist and was advised to undergo Magnetic Resonance Imaging (MRI) and Computed Tomographic (CT) scans. The MRI scan demonstrated a vascular loop closely abutting right IX cranial nerve. From the MRI results, condition was diagnosed as Glossopharyngeal neuralgia (GPN) due to microvascular compression. He was then prescribed T. oxetol (oxcarbazepine) 150 mg 1-0-1, which provided a temporary relief for the patient. So, he was advised to undergo neurosurgery.

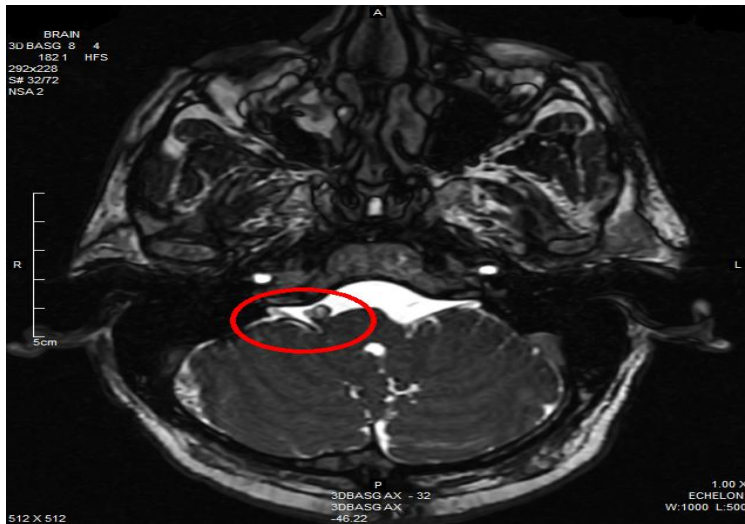
**DISCUSSION**

Weisenberg syndrome was first described by Weisenberg in 1910 in a 36-year old male patient with cerebellopontine angle tumour[1]. Some studies demonstrated that the incidence rate of this rare pain syndrome to be approximately 0.7 per 100,000 population per year[2][9]. The disease generally occurs in adults and has its onset usually after an age of 40-years. The incidence of disease is slightly higher in men than women[3][4][8].

Most of the cases of GPN occur in isolation (idiopathic). The major etiologies for GPN includes microvascular compression (compression of glossopharyngeal nerve by a nearby blood vessel,

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usually the posterior inferior cerebellar artery), multiple sclerosis, tumour of brain and neck, Varicella – Zoster infections and Eagle’s syndrome (nerve compression by elongated styloid process of skull). The secondary causes of GPN involves Paget’s disease, Arnold - Chiari malformation and neck trauma with foreign body impaction[5].



**Figure 1:** MRI of brain (posterior fossa) reveals a left sided dominant circulation with prominent tortuous vertebral artery and branches causing compression of the neural bundle entering the jugular foramen. The offending vessel is not clearly seen.

The International Headache Society classified GPN into two categories i.e. Classic and Symptomatic. Classic GPN is presented as severe transient stabbing pain experienced in the ear, base of the tongue, tonsillar fossa or beneath the angle of jaw. It can be exaggerated on swallowing, laughing or chewing. But, Symptomatic GPN involves the added presence of an aching pain that usually persists between the attacks. The pain due to GPN is felt in the distributions of the auricular and pharyngeal branches of glossopharyngeal (IXth cranial nerve) and vagus (Xth cranial nerve) nerves[5][8][11].

The diagnosis is made on clinical grounds alone. GPN can be distinguished from trigeminal neuralgia based upon the location of pain. In GPN, the application of a local anaesthetic (like lidocaine) to the throat temporarily eliminates spontaneous or evoked pain[7]. High resolution MRI scan is beneficial if the offending compressing artery is PICA (posterior inferior cerebellar artery), because it will show a loop formation at the supraolivary fossa (middle portion of cerebellopontine-medullary angle) in MRI. In case the offending compressing artery is anterior inferior cerebellar artery (AICA), it is very difficult to diagnose GPN before surgery[6].

The first line treatment for GPN is pharmacological. Anti-seizure drugs such as oxcarbazepine (300 - 1200 mg/day) and carbamazepine (400 - 1200 mg/day) are the primary choices. But, other drugs like gabapentin, pregabalin and some anti-depressant medications are also used[1][7]. But, when the pain cannot be managed effectively by medications, surgical intervention is indicated. Microvascular decompression (MVD) is the first surgical option as it has highest initial and long-term success rates. In patients who cannot tolerate such an operation due to age restrictions or other medical problems may undergo less invasive surgeries like Gamma knife stereotactic radiosurgery[7]. Vagus nerve rhizotomy is performed in conditions where vascular conflict is not evident[10].

In this case, our patient had Classic GPN which was inferred from his symptoms. The major reason for his condition was found to be compression of glossopharyngeal nerve by tortuous vertebral artery and branches. The patient was subjected to microvascular decompression and glossopharyngeal neurectomy. He had satisfactory relief from pain and was discharged.

## CONCLUSION

In summary, Weisenberg syndrome is rare compared to other facial pain syndromes. No single procedure is best for everyone and each procedure varies in its effectiveness versus side effects. The cases of Weisenberg syndrome must be thoroughly assessed and diagnosed by vascular imaging techniques like MRI – scan and CT - angiogram to determine the underlying cause and choose appropriate treatment regimen.

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