When intra-operative exploration is the only option, severe medically refractory trigeminal neuralgia

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ABSTRACT

Introduction: Patients with trigeminal neuralgia (TN) are usually treated on an outpatient basis, and symptoms can be controlled using various medical therapeutic options. We present a case of severe TN with an acute on chronic flare, which was now refractory to a wide variety of medical options, had a prolonged inpatient hospitalization, and ultimately required surgery with excellent results.

Case: A 54-year-old Hispanic male was admitted with severe left-sided TN. His symptoms gradually became more pronounced and frequent to a point where he was unable to have a meaningful quality of life. A trial of gabapentin, phenytoin, opioids, and NSAIDs had also been unsuccessful before this hospitalization. He had three ER visits before he was finally hospitalized with intractable pain and unfortunately had begun to have suicidal thoughts. Various therapeutic interventions were tried, including escalating doses of opioids and local nerve blocks, but all non-surgical options failed to provide relief. Neurosurgical route was eventually approached, and patient underwent left retro mastoid suboccipital craniectomy. Intraoperatively, the left superior cerebellar artery was found to adhere to the inferior ventral aspect of the left trigeminal nerve root entry zone. Upon decompression patient's symptoms resolved dramatically. Interestingly this vascular compression was not seen on multiple prior brain imaging.

Conclusion: TN can severely affect someone's quality of life. It often leads to severe anxiety and depression. Our case represents the importance of proceeding towards surgical options sooner rather than later. An early multi-disciplinary approach is warranted.

1. Introduction

Trigeminal neuralgia (TN) is one of the most common causes of facial pain with an annual incidence ranging from 4 to 13 per 100,000 in the USA [1]. Patients with TN are usually treated on an outpatient basis, and symptoms can be managed with various medical therapeutic options. It is a condition that is characterized by recurrent episodes of neuropathic pain and is usually abrupt in onset and termination. Pain occurs along with the distribution of one or more divisions of the 5th cranial nerve.

The trigeminal nerve has three major divisions: ophthalmic V1, maxillary V2, and mandibular V3. Etiology can be divided into idiopathic, primary, or secondary causes. A primary cause is described as a neurovascular compression in the trigeminal root entry zone, resulting in nerve root atrophy or displacement. Compression and distortion of the trigeminal nerve by a tortuous and elongated superior cerebellar artery are seen in up to 80–90% of cases [2]. Secondary causes include underlying tumors like acoustic neuroma, meningioma, aneurysm, or AV malformation. A correlation between TN and multiple sclerosis has also been reported in the literature [3].

Typically pain is acute in onset, lasts briefly and is usually severe in intensity. In approximately 60% of cases, only one branch (maxillary or mandibular) is involved, whereas in about 35%, both are involved. In less than 4% patients, only ophthalmic nerve is affected. Pain sometimes can be accompanied by facial muscle spasms which were very apparent in our case described here [4].

There is strong evidence to believe that carbamazepine or oxcarbazepine should be offered as first-line therapy for TN. There is little evidence to support other alternatives like baclofen, lamotrigine, and NSAIDs. Surgical interventions include microvascular decompression, rhizotomy, and gamma knife radiosurgery. This report presents a case of severe TN requiring inpatient hospitalization who was refractory to medical therapy and eventually required surgical intervention.
2. Case

This is regarding a 54-year-old Hispanic male with no significant past medical or surgical history other than left-sided TN. He first developed this disease in 2019, and at that point was started on carbamazepine 400 mg twice daily. This worked reasonably well to control his neuralgic pain and he was able to continue doing his job until later part of 2020, when he started seeing his primary care physician more frequently. His symptoms gradually became more pronounced and frequent. He was now no longer able to work his usual job and had severe frequent intermittent episodes of left-sided facial pain. He subsequently had minimal oral intake and was barely able to sleep enough. By this time, his PCP had maximized his dose of carbamazepine but it failed to achieve any improvement. An outpatient trial of gabapentin, phenytoin, opioids, and NSAIDs had also been unsuccessful. In a span of 10 days, he had three ER visits before he was finally hospitalized with intractable pain and subsequent suicidal ideations. Family history of chiari malformation was reported in one of the siblings. His physical exam on initial presentation included normal vital signs. Neurological examination was pertinent with hyperesthesia reported along with the left lower V1 and throughout the entire left V2 cranial nerve distribution.

Many therapeutic interventions were tried, including escalating doses of intravenous opioids, fosphenytoin loads and subcutaneous triptans but with minimal relief. He eventually required sedation with benzodiazepines to control pain and severe suicidal thoughts. After five days of different therapeutic interventions with no significant benefit, patient was transferred to a tertiary level care facility under the care of neurology and internal medicine. Upon arrival to the facility, a trial of ketamine infusion was attempted immediately but had to be stopped due to development of hallucinations. A trigeminal nerve block was attempted but gave minimal and transient relief only.

In the interim patient had multiple imaging studies conducted, including Brain MRI and CT angiogram none of which truly identified an etiology. Given refractoriness to medical interventions, neurosurgical team was approached. Patient eventually underwent left retro mastoid suboccipital craniectomy and intraoperatively the left superior cerebellar artery was found to adhere to the inferior ventral aspect of the left trigeminal root entry zone. Lysis of arachnoid adhesions was performed to separate this vessel and relocation to a more ventral lateral position, thereby achieving microvascular decompression. Postoperatively, patient was noted to have a dramatic improvement in his left-sided neuralgic pain and had discomfort over the surgical site only. He was monitored in the intensive care unit for about 24 hours and was discharged to home. A brief follow-up was done over the phone with the patient into four months following surgery. To this point, his symptoms have not recurred.

3. Discussion

In 2008, the American Academy of Neurology (AAN) and the European Federation of Neurological Societies (EFNS) combined developed guidelines to help providers in the management of TN, by addressing questions related to its diagnosis, medical, and surgical treatment. There is insufficient evidence to suggest when surgical intervention should be offered however some TN experts recommend early surgical referral to patients who fail to respond to first-line therapy. A cross-sectional survey was conducted at a specialized center in England which found that as high as 40% of the 129 respondents eventually required surgical intervention for TN. However, it is important to understand that despite of the higher chances of requiring surgery, it does not come without possible major complications. Hence, surgery is reserved for people who still experience debilitating pain despite the best medical management [5].

Our patient fell in that category where he was now refractory to wide medical therapeutic options. Over the course of few weeks, symptoms in this case progressively became worse but fortunately resolved almost completely when the nerve was surgically decompressed. Over time, TN is often known to increase the risk for developing depression, anxiety, and sleep disorders [6]. This was very apparent in this patient’s course as well. His persistent symptoms made him incapable of performing his job and were pointing him towards permanent disability, thereby causing significant depression to a point where he started having severe suicidal ideations.

Another important thing to highlight from this case is that there were several brain imaging studies performed, including MRI and CT scans with angiograms but no obvious vascular loop compression was seen preoperatively. This is not an uncommon finding and is somewhat expected. For patients with TN without non-trigeminal neurological symptoms, routine neuroimaging possibly identifies a cause in up to 15% of patients only [7,8].

4. Conclusion

Trigeminal neuralgia can severely affect someone’s quality of life. It often leads to severe anxiety and depression. Our case represents the importance of
proceeding towards surgical options sooner rather than later. An early multi-disciplinary approach is warranted.

**Disclosure statement**

No potential conflict of interest was reported by the author(s).

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