A Case of Occipital Neuralgia

Abstract

Occipital Neuralgia (ON) is a rare form of headache characterised by paroxysms of stabbing pain in the distribution of the Greater or Lesser Occipital Nerves. Because of difficulty in diagnosis, no reliable prevalence or incidence data is available. It is diagnosed both clinically and by response to blockade of the relevant occipital nerve with local anaesthetic. The condition can be very hard to manage and life changing for a patient.

Conservative management including physiotherapy and pharmacotherapy is often unsatisfactory and nerve blocks have transient effects (often under 3 months). Surgical and Neuromodulatory solutions have the potential permanence but also potential for significant complications.

The case presented is that of a young female with Occipital Neuralgia, likely to be secondary to previous surgical intervention, that highlights the management difficulties of this condition. Her Neuralgiform headaches were refactory to simple treatments and Occipital Nerve blocks led to temporary relief. This is significantly prolonged by the addition of pulsed radiofrequency to the Greater Occipital Nerve. A review of the management options includes conservative and interventional approaches to this complex condition.
Case Presentation

A 35 old female had been diagnosed with a right-sided acoustic neuroma after presenting with gradually increasing dizziness and unilateral hearing loss. She had a microsurgical resection via a trans-labyrinthine approach and at the same time had a bone anchored hearing aid implanted. Surgery was uneventful.

Although the surgery was successful at removing the tumour, over the next six months she began to complain of a persistent neck pain and headache on the operated (right) side. Subsequent MRI & CT scans revealed no evidence of tumour recurrence or any other abnormality. Treatment of this pain was unsuccessful with simple analgesics (Ibuprofen & Paracetamol) so she was referred to the Chronic Pain team by her neurosurgeon.

She had no other significant medical history and was otherwise well with no allergies and on no regular medication except: Paracetamol 1g QDS & Ibuprofen 400mg TDS, which did not help her headache.

She described her pain as having two components. The first was a deep aching constant pain at the back of her neck made worse by movement. The second pain was much more severe with a pulsating or occasionally burning character originating from the right neck around the level of C2, paravertebrally and radiating to the right occipital scalp. Occasionally it then radiated into her anterior scalp and just behind her eyes. She stated that the second pain was the most problematic for her, although it was intermittent, it could occur up to ten
times a day lasting from minutes to hours. Episodes of severe pain could come on spontaneously but were sometimes evoked by touching the back of her head or moving her neck.

Her baseline Numerical Rating Score was (NRS) 4/10 in intensity. This increased to up to 8/10 in intensity when the lancinating episodes of pain occurred.

She continued to work full time once she had recovered from her initial surgery and had no evidence of catastrophisation, depression or anxiety. Her sleep was mildly affected due to pressure of the pillow leading to headaches but she had adapted by sleeping on her other side.

Clinical Examination revealed no abnormal neurology or evidence of infection. Deep palpation lateral to the occipital condyle on the superior nuchal line provoked severe spasms of pain as did neck hyperextension. A provisional diagnosis of occipital neuralgia was made. This was thought to be likely secondary to her previous surgical intervention as surgery for acoustic neuroma has been linked to secondary occipital neuralgia³.
**Case Management**

Initially a non-interventional approach was adopted, but neck physiotherapy resulted in no improvement in symptoms. Anti-neuropathic pain medication was tried next; Nortriptyline had no benefit and sedation side effects with the Gabapentinoids resulted in cessation of treatment.

Greater Occipital Nerve Blocks were performed aseptically via a landmark approach in the sitting positioning with neck flexed. A 27g needle was inserted to bone approximately 1/3 of the distance between the occiput and mastoid along the nuchal line medial to arterial pulsation. Lignocaine 1% (1.5ml) and Triamcinolone (20mg) was injected. Within five minute her VAS had fallen to Zero, this confirmed the diagnosis as occipital neuralgia. Unfortunately the effects of these were, temporary lasting only 4-6 weeks before her symptoms returned.

Peripheral Electrical Nerve Stimulation (PENS) of the Greater Occipital Nerve was performed by the Neurosurgical team. This led to pain cessation but only for around 48 hours and was found very uncomfortable by the patient.

To prolong the duration of analgesia from the greater occipital nerve blocks, Pulsed Radiofrequency of the Greater Occipital Nerve was performed. This was applied to the greater occipital nerve in addition to injection with lignocaine and triamcinolone.
The skin was anaesthetised with 1% lignocaine. A 20g insulated needle with 10 mm exposed tip was inserted via the same landmarks described above. The needle was inserted obliquely so as to increase the length of the stimulating tip in proximity to the nerve. Pulsed Radiofrequency current was applied for two intervals of 120 seconds at 42°C (2Hz 20ms pulses).

As with the steroid and local anaesthetic injection, her VAS dropped to zero immediately post procedure. Significantly, the addition of Pulsed Radiofrequency increased the duration of her analgesia to 10-12 weeks.

Though this was a significant improvement on her previous pain control, the patient felt that needing to attend hospital for invasive procedures every three months for the rest of her life would not be ideal. As a result she is now awaiting either an Occipital Nerve Stimulator or a Greater Occipital Neurectomy for a permanent solution to her intractable occipital pain.
**Discussion**

Occipital neuralgia is a rare form of headache characterised by pain in the distribution of the greater or lesser occipital nerves. These derive from the dorsal rami of the second and third cervical nerve roots respectively and innervate the posterior scalp. The Greater Occipital Nerve is more frequently the culprit than the lesser, 90% vs 10% of cases. In 8.7% of cases both nerves are involved.

The Greater Occipital nerve origin is lateral to the atlantoaxial joint from the medial branch of C2. It ascends superficially to Rectus Capitis Posterior Major and pierces Semispinalis Capitis. Passing above an aponeurotic sling (from Trapezius and Sternocleidomastoid) it becomes subcutaneous and then lies immediately medial to the occipital artery where it is accessible for superficial blockade along the Superior Nuchal line between the Occipital Condyle and Mastoid Process.
Figure 1: Anatomy of the Occipital Nerves

(Image taken from Science Direct)
The International Classification of Headache Disorders (ICHD) has three criteria for the diagnosis of occipital neuralgia to be confirmed⁵.

1) Paroxysmal spasms of pain +/- persistent aching pain in the distribution of the greater or lesser occipital nerves
2) Tenderness over the affected nerve
3) Temporary relief with local anaesthetic block

Other commonly associated symptoms are ocular pain, visual disturbance, tinnitus, dizziness and nausea. These are thought to be due to the interrelation of the occipital, the trigeminal and sympathetic nerves.

It is important to make sure the diagnosis is not confused with other more common differential diagnoses of occipital neck pain and cervicogenic headache including:

- Migraine
- Tender Trapezius / Myofascial Trigger points
- Cervical Facetogenic Pain
- Atlanto-Axial Joint Pain
Occipital Neuralgia can be idiopathic but is often secondary to a structural nerve injury. It is usually relatively easy to distinguish between primary or secondary ON based on the patient history and cervical spine MRI. Examples of secondary causes include:

- Cervical Spine degenerative changes resulting in C2 / C3 foraminal narrowing and nerve root compression. (more accurately this is a C2 radiculopathy than ON)
- Trauma (including direct nerve trauma and hyperextension/whiplash injury)
- Surgery
- Vascular Compression
- Malignancy affecting C2/3 Roots
Management of Occipital Neuralgia

Management of any pain condition involves a careful history and examination to exclude other differential diagnoses and a multimodal, biopsychosocial approach. Because of its relative rarity, evidence for most treatments for occipital neuralgia is based on small samples and equates to Level III-IV evidence.

Table 1: Management of Occipital Neuralgia

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Non Invasive Approaches

Conservative approaches to the management of ON include physiotherapy, massage and acupuncture. The objective is to reduce any muscle spam that is leading to or worsening nerve entrapment. Evidence of any benefit is poor.

Anti neuropathic pain drugs are widely used for ON but there have been no clinical trials specifically looking at the efficacy of these drugs in this situation. Drugs widely used include the Tricyclic Antidepressants and the Gabapentinoids.
Invasive Approaches

The superficial position of the occipital nerves and the focal nature of the condition are very conducive to an interventional approach to this condition. As described above, a positive response to local anaesthetic blockade of the Occipital nerves is mandatory to diagnose the condition.

Greater Occipital Nerve Block

Greater Occipital nerve blocks have long been performed for occipital neuralgia as well as for cervicogenic headache and migraine. Under sterile technique, a needle is inserted subcutaneously until bone is reached approximately 1/3 of the distance between the occipital protruberance and the mastoid along the superior nuchal line\(^4\). If palpable, this should be just medial to the arterial pulsation. After careful aspiration to exclude arterial puncture, local anaesthetic with depot steroid (triamcinolone or methylprednisolone 20-40mg) is injected in a fan-like distribution; a total of 1-3ml is injected.

Side effects are rare but include - infection, bleeding or bruising and intravascular administration of local anaesthetic. There have also been case reports of localised alopecia\(^6\). The local anaesthetic leads to an immediate but transient improvement in symptoms but the steroid will not have a significant benefit until several days have passed. The mechanism of analgesia from the steroid is thought to be by reducing inflammation within and around the nerve leading to a reduction in nerve compression and thus nerve hyperexcitability.
Overall this leads to a reduction in muscle spasm and headache. Multiple studies have shown that nerve blocks help with ON but their effects are usually transient.

**Pulsed Radiofrequency of the Occipital Nerve**

Pulsed Radiofrequency (PRF) has been used to prolong the duration of Occipital Nerve Blocks. The mechanism of pulsed radiofrequency is not fully understood. Radiofrequency is applied at two (20ms) pulses per second, this prevents excessive heat generation, keeping the temperature below 42°C. This prevents neuroablation and tissue damage. Despite the lack of visible damage, a large electrical field is conducted focally into the target tissue and this is thought to mediate its effects. These electric fields affect the transmembrane potentials of the nerves leading to long-term depression of signalling. Structural changes can be seen in nerves post PRF including changes in both cellular and organelle membranes, mitochondria, microtubules and microfilaments. Pulsed RF has been used widely for neuropathic pain at both peripheral nerves and dorsal root ganglia.

PRF has been done to the C2 dorsal root ganglia for ON but this has potentially severe complications including; subarachnoid haemorrhage, vertebral artery puncture and nerve injury and has no greater evidence of efficacy than peripheral application of PRF.
PRF has been applied the Greater Occipital Nerve peripherally at the Superior Nuchal Line as describe in this case report. Choi et al described a case series of 10 where PRF to the Greater Occipital Nerve led to complete pain relief in 9 and substantial pain relief in 1 for a period of at least 6 months\textsuperscript{7}. Vanaelderden et al also demonstrated significant improvements in analgesia even after 6 months in 52.6\% of patients after pulsed RF of the Greater Occipital Nerve\textsuperscript{8}.

A recent study by Huang et al on 102 patients having PRF for ON found that 52 obtained >50\% pain relief for more than 3 months\textsuperscript{9}. A retrospective analysis was then performed to find factors associated with a positive benefit of PRF. Factors that were associated with positive benefit include a low block volume and ON secondary to initial trauma. Factors associated with a poor response include pain spreading anteriorly and evidence of secondary gain.

**Occipital Nerve Stimulation\textsuperscript{10}**

Occipital nerve stimulation (ONS) is viewed by many as the gold standard for the management of occipital neuralgia ever since its first use by Weiner and Reed in 1999\textsuperscript{11}. This report on 13 patients with ON showed all patients who had a trial of occipital stimulation going on to have permanent implantation and all getting good or excellent pain relief even after 1 \(\frac{1}{2}\) years follow up.
ONS is a type of peripheral neuromodulation\textsuperscript{1,2,13}. One or two electrodes are inserted perpendicular to the course of the greater and lesser occipital nerves usually at the level of C1. A positive response to a GONB with local anaesthetic is not a screening test for response to ONS. Instead a percutaneous trial of stimulation is performed and if there is a good response then a permanent system is implanted. The permanent system can either be inserted percutaneously or surgically. An implantable pulse generator is implanted into a subcutaneous pocket and connected via a tunneled wire.

Good results have been seen with ON, migraine and cluster headache. Multiple case series and case reports have been done for stimulation in ON and results are perhaps unsurprisingly much better than for other headache types\textsuperscript{10}. So far all studies performed have shown an overwhelmingly positive, long-term benefit but it should be noted that due to its invasive nature, there has been no blinded studies.

Other disadvantages are that this is an expensive technique, costing around £20,000 and has a substantial complication rate with high rates of lead migration (~25%) and disconnection because of neck movement. Other complications include infection (~5%) and battery failure. Overall revision rate in the literature is ~10-60%\textsuperscript{14}. Surgical implantation and the use of a paddle, rather than a cylindrical wire electrode has been associated with lower rates of migration and disconnection.
Figure 2: Paddle placement for Bilateral Occipital Nerve Stimulation

Taken from Ref 10
Surgical Techniques

Due to the temporary nature of GONBs and the complications and costs of stimulators, many surgical approaches have been applied to achieve permanent analgesia. These include: neurolysis, neurectomy, C2 Gangliotomy or Gangliectomy, & C2-C3 Rhizotomy or Radiofrequency denervation. These have often been done on very small patient numbers with variable results. They can be separated into nerve release procedures and neuro-ablative procedures.

There are significant concerns about ablating such a large sensory nerve due to the risk of deafferentation pain. A review of neurectomy techniques for ON showed that although 70% of patients had pain relief at 18 month follow up, 30% had substantial side effects including: unpleasant dysesthesias, hyperalgesia and neuroma formation. However a study, by Ducic et al on patients with ON secondary to acoustic neuroma surgery found that 6 out of 7 patients who had Greater Occipital Nerve excision had 80% reduction in quality of life at 32 months.

Ducic et al published results on a series of 190 patients with ON who underwent surgical greater occipital nerve release. 80 percent of patients had significant pain relief at 12 month follow up although only 43% had complete analgesia.
Conclusions

Occipital Neuralgia is a rare form of headache that is often refactory to simple medical management. Due to its rarity there is little data on its accurate incidence and prevalence and most evidence on its management would be classed as level III-IV\textsuperscript{13}. Also this means that there is no agreed gold standard patient management pathway.

Occipital Nerve blocks have been shown to be effective and are mandatory for diagnosis but are short acting and Pulsed Radiofrequency may be a low risk adjunct with the potential to significantly increase the duration of these blocks. Further research needs to be performed to formalise this techniques role and efficacy and calculate the optimum technique and duration of stimulation.

When nerve blocks are too short lasting, permanent solutions such as Occipital Nerve Stimulators are the most effective long-term solution but are expensive with a significant risk of complications. Further research on reducing the lead migration rate would substantially improve the cost effectiveness of stimulation. If achieved, this would reduce the need for many patients to undergo surgical neurolytic procedures with the substantial risk of longstanding deafferentation pain.
The case described outlines the classic difficulties of managing these complex patients and the gradual migration from a non-interventional to highly invasive management of this condition. In this case, the patient despite obtaining a longer duration of pain relief with pulsed radiofrequency to the Greater Occipital Nerve is now awaiting surgical ligation of her Greater Occipital Nerve.
References

1) Vanelderen P. et al Occipital Neuralgia Pain Practice Vol 10, Issue 2, pp 137–144 2010


12) Neuromodulation in Primary Headache. IASP Clinical Updates Vol XX issue 5 2012


