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ORIGINAL ARTICLE

Case series: non vascular considerations in trigeminal neuralgia

Sathesh Balasundram · Stefano Cotrufo · Colin Liew

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Abstract An abnormal vascular course of the superior cerebellar artery is often cited as the cause for trigeminal neuralgia. However, among patients with TN-like symptoms, 6% to 16% are variously reported to have intracranial tumours. Aneurysms, tumours, or other lesions may impinge or irritate the trigeminal nerve along its course. Uncommonly, an area of demyelination from multiple sclerosis may be the precipitant. We would like to present a series of unusual lesions, all of which initially presented with neuralgic-like symptoms and were refractory to treatment. Collated case series with photographs and imaging are reviewed in this paper. Discussion of case presentation and management are done for evaluation. A wide range of other compressive lesions can cause trigeminal neuralgia. This paper illustrates the clinical presentation of atypical trigeminal neuralgia and emphasises the value of diagnostic imaging in trigeminal neuralgia patient. Suggested algorithm for management of trigeminal neuralgia

Keywords Trigeminal neuralgia · Neuropathy · Intracranial tumour

Abbreviation Key

TN	Trigeminal neuralgia
MS	Multiple sclerosis
IASP	The International Association for the Study of Pain
IHS	International Headache Society
MRI	Magnetic resonance imaging
CT	Computerised tomography
CTA	Computerised tomography angiogram

S. Balasundram (✉)
Oral and Maxillofacial Surgery Unit, Eastman Dental Hospital,
University College London Hospital,
London, UK
e-mail: satbala@yahoo.com

S. Cotrufo · C. Liew
Department of Oral and Maxillofacial Surgery, University College London Hospital,
London, UK

Introduction

Trigeminal neuralgia is characterised by sharp transient attacks of pain affecting the dermatomal distribution of the trigeminal nerve. It is often described as paroxysmal, stabbing, lancinating, and electric shock like [1]. Trigeminal neuralgia (TN) may be an extremely disabling illness, capable of causing such suffering that it is sometimes referred to as the 'suicide disease'.

The first known description of trigeminal neuralgia, or a similar condition, was written in the second century AD by Aretaeus of Cappadocia, a contemporary of Galen. Also known for his descriptions of migraine, he makes reference to a pain in which 'spasm and distortion of the countenance take place' [2]. Jujani, an eleventh century Arab physician, mentions unilateral facial pain causing spasms and anxiety in his writings. Interestingly, he suggests that the cause of the pain is 'the proximity of the artery to the nerve' [3]. An example of early interventional treatment is that by Locke in 1677, who applied sulphuric acid to the face of the Duchess of Northumberland in an attempt to treat her trigeminal neuralgia [4].

The first full account of TN was published in 1773 when John Fothergill presented a paper to the Medical Society of London. He described the typical features of the condition in detail, including paroxysms of unilateral facial pain, evoked by eating or speaking or touch, starting and ending

abruptly, and associated with anxiety [2]. Some time earlier, Nicolaus André had used the term 'tic douloureux' to describe what he thought was a new clinical entity [3].

Sporadic observations later in the eighteenth and nineteenth century by Pujol, Chapman, and Tiffany helped to complete the clinical picture and differentiate TN from common facial pain conditions such as toothache. In the early twentieth century, Oppenheim alluded to an association between multiple sclerosis (MS) and TN, and Patrick commented on its familial incidence [5].

The International Association for the Study of Pain (IASP) and International Headache Society (IHS) have suggested their own diagnostic criteria for TN [6, 7]. These are remarkably similar and highlight the sudden, explosive nature of the pain. In further descriptions of the condition, both classifications allude to vascular compression, MS, and tumours as known aetiological causes. The IASP classification makes a distinction between TN (including MS) and secondary neuralgias (caused by structural lesions and injuries, but not including MS), while IHS separates idiopathic TN from the 'symptomatic form' depending on the presence of a structural lesion; it is not quite clear if vascular compression qualifies as such. Neither approach includes reference to variant forms of TN, which satisfy the diagnostic criteria but display additional features as well.

Classical trigeminal neuralgia has an annual incidence of ~3–27 per 100,000. The incidence increases with age and the condition has a female predisposition [1, 8].

As Fothergill observed, the symptoms of trigeminal neuralgia are quite distinct. The electric, lancinating pain has an abrupt onset and termination, lasts a few seconds, and can be so intense that the patient often winces in a tic-like fashion. The pain is limited to the distributions of the trigeminal nerve. Cutaneous stimuli within specific 'trigger zones' may precipitate a pain attack, leaving some patients unable to chew, drink, shave, or brush their teeth. Occasionally, the site of pain is remote to the site of stimulation. Some patients will have a brief refractory period following an attack, during which subsequent cutaneous stimuli will not trigger an episode. There is often diurnal variation in the pain attacks, with frequent morning exacerbations. This may be due to either 'wearing-off' of medications or to concentrated periods of facial stimulation accompanying morning activities. Pain attacks are characteristically absent during sleep. Symptoms often occur in bouts lasting weeks to months, and initially there are periods of spontaneous remission [9].

Among patients with TN-like symptoms, 6% to 16% are variously reported to have secondary neuralgias, most commonly due to intracranial tumours. The aim of this paper is to present a series of cases with secondary TN. We report here a series of seven cases with patients presenting with non-vascular-related trigeminal neuralgia.-

Case series

Case 1

An 82-year-old woman presented with left-sided trigeminal neuralgia. An MRI scan showed left-sided cerebellar pontine angle epidermoid cyst extending anteriorly into Meckel's cave and compressing the trigeminal nerve. There was no mass effect and her symptoms are well controlled (Fig. 1) with carbamazepine. She is under surveillance.

Case 2

A 68-year-old woman had a 3-year history of paraesthesia of her left face which progressed to pain. She indicated a pain map of maxillary and mandibular divisions of the trigeminal nerve. A mass was palpable medial to the left ramus of the mandible. CT and CTA showed a well-defined mass in the left medial pterygoid area which extended to the base of skull with several branches of the maxillary artery draped over it (Fig. 2). Features suggested an osteochondroma. Symptoms were controlled with conventional treatment. The patient declined surgical intervention.

Case 3

A 23-year-old patient presented with anaesthesia dolorosa in the maxillary division with no focal signs. MRI showed a large skull base mass occupying the right infratemporal fossa extending from the pterygoid region, through the parapharyngeal space and into the posterior skull base (Fig. 3). The lesion appeared to extend from the trigeminal ganglion. Tissue was obtained via a transantral approach.

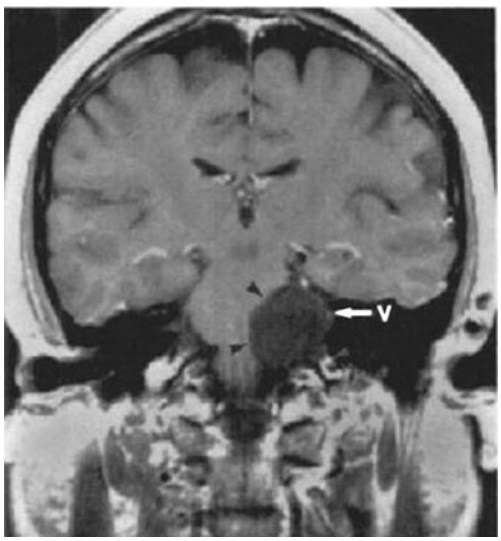

Fig. 1 MRI showing left cerebellar pontine angle cyst

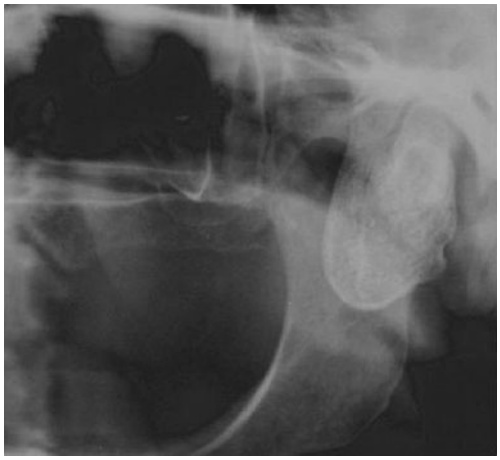

Fig. 2 Sectional OPG showing mass over left ramus of mandible

This confirmed the provisional diagnosis of a trigeminal nerve schwannoma. Plans to resect the tumour via a combined antero-lateral and transzygomatic approach were declined.

Case 4

A 54-year-old dental practitioner presented with unremitting pain in the distribution of his right mental nerve. He had made a self-diagnosis of trigeminal neuralgia and had been treating himself with escalating doses of carbamazepine with little relief. An orthopantomogram showed a multilocular lesion in angle of mandible which was diagnosed as an ameloblastoma (Fig. 4). He subsequently had a mandibular resection and reconstruction.

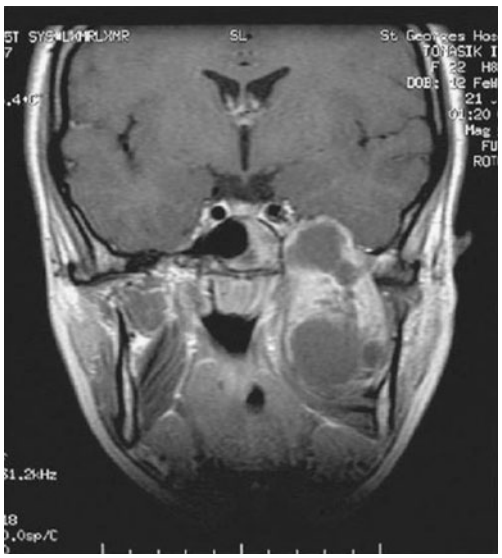

Fig. 3 MRI showing large skull base tumour extending from the pterygoid region to the posterior skull base

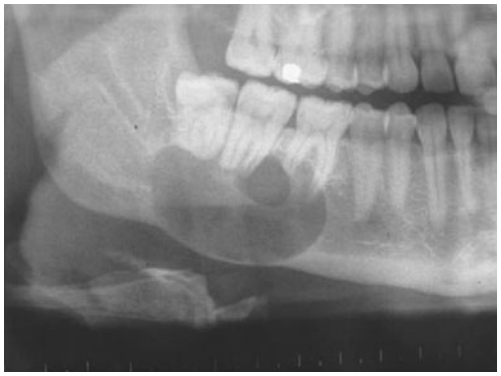

Fig. 4 OPG showing large multilocular lesion over angle of mandible

Case 5

A 46 year old presented with a numb lip with a deficit defined by the mandibular branch of the trigeminal nerve. MRI showed a vestibular schwannoma in the left cerebellapontine angle (Fig. 5). He had a posterior fossa approach for excision of the tumour.

Case 6

An 81-year-old woman with a rapidly enlarging right-sided parotid lump developed trigeminal neuralgia, and then subsequently multiple cranial nerve neuropathies (V, VI, VII, VIII). An MRI scan showed a large lesion with a finger of tumour extending intracranially through the foramen ovale (Fig. 6). An open biopsy of the parotid was performed. The diagnosis was of a large B cell lymphoma.

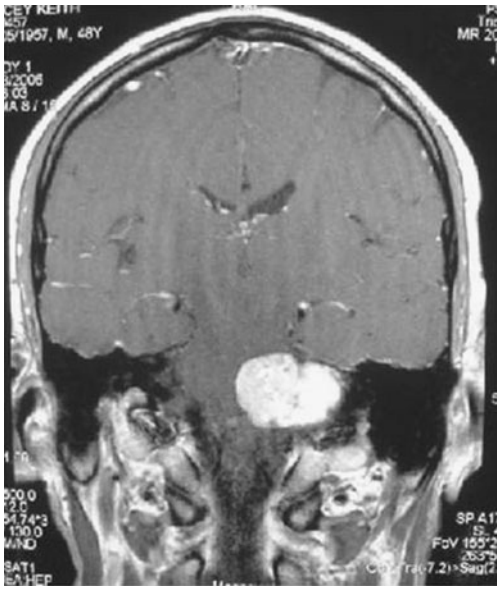

Fig. 5 MRI showing coronal view of tumour in left cerebellar pontine angle

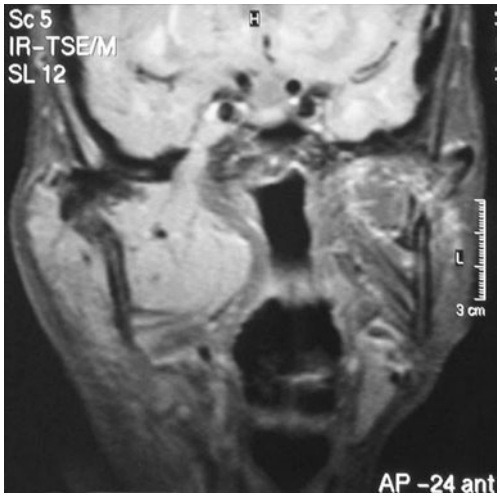

Fig. 6 Coronal MRI view of a large tumour with a finger of extension via foramen ovale

Case 7

A 52-year-old gentleman presented with sensory deficit of his left V_3 and symptoms consistent with left trigeminal neuralgia. No specific triggers were noted and he had normal taste and facial movements. The patient was diagnosed with trigeminal autonomic cephalgia although no definite cranial autonomic features were identified.

He presented a year later with a 6-month history of the cervical lymphadenopathy. Ultrasound scans revealed a left submandibular lump corresponding to the left level 1b lymph nodes. An incidental abnormal hypoechoic soft tissue directly adjacent to the buccal aspect of the left side of the mandible was noted with marked widening of the left mental foramen. MRI scans showed a rim of abnormal soft tissue over the buccal aspect of the left side of the mandible and in the mental region. The involvement of the medullary cavity of the left side of the mandible with widening of the left mental foramen is shown on CT scan (Fig. 7). The biopsy results reported this as a hairy cell leukaemia. The patient has been referred to the lymphoma services for further management.

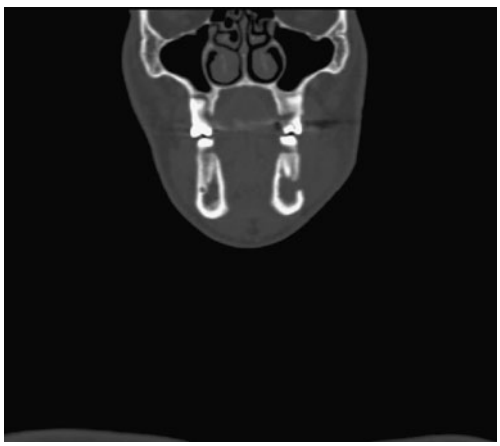

Fig. 7 CT scan revealing widening of left mental foramen

Discussion

Neuralgia of the trigeminal nerve can involve its course from its nuclei in the brain stem to its peripheral branches. Dysfunction may be a consequence of supranuclear, nuclear, or infranuclear disease. This paper reports a series of trigeminal neuralgia secondary to intracranial and extracranial tumours.

Much the commonest cause of trigeminal neuralgia is focal compression of the trigeminal nerve root, close to its point of entry into the pons, by an aberrant loop of artery or vein. This was first recognised as a cause of trigeminal neuralgia by Jannetta (1967) [10] and is now thought to account for 80–90% of cases.

Trigeminal neuralgia is a well-recognised complication of multiple sclerosis. Typically, a plaque of demyelination encompasses the root entry zone of the trigeminal nerve in the pons. Rarely, patients with peripheral nerve demyelination due to Charcot–Marie–Tooth disease develop trigeminal neuralgia [11]. MS is seen in 2–4% of patients with TN. Conversely, TN is diagnosed in 1–5% of patients with MS. In a small proportion of patients with MS, TN is the first manifestation of the disease. These patients are younger than the TN population as a whole and their neuralgia is more frequently bilateral. A latent demyelinating disease should be considered in young patients with TGN and appropriate diagnostic tests performed, as disease-modifying treatment for MS is emerging [9].

Other, rare causes include infiltration of the nerve root, gasserian ganglion, or nerve by a tumour or amyloid, and small infarcts or angiomas in the pons or medulla. Rarely, trigeminal neuralgia results from vascular compression of the nerve root by a saccular aneurysm [12] or an arteriovenous malformation [13].

A wide range of other compressive lesions can cause trigeminal neuralgia. These include vestibular schwannomas [14], meningiomas [15], epidermoid cysts [16], and various other cysts and tumours [17–20]. In several reported cases, the neuralgia was contralateral to the side of the mass lesion [15, 18, 19]. Compression of the trigeminal nerve root may be mediated by the tumour itself, by an interposed blood vessel or by distortion of the contents of the posterior fossa with displacement of the nerve root against a blood vessel or the skull base [8]. Once all of these possibilities have been excluded, there remains a small proportion of patients in whom the aetiology is undetermined [8].

Chmielewska and Kamiński (2003) [21] reported of a patient with first division trigeminalgia without any

neurological loss and with normal laryngological, ophthalmological, neurological, and stomatological examinations, as well as imaging. After 1 year, a MRI of the orbit revealed a pathological mass in its apex with a connection to the superior orbital fissure that was confirmed to be leiomyosarcoma. It is interesting to note in this report that trigeminal neuralgia presented as a preceding symptom to the tumour. This was similar to cases 2, 4, and 7 in our report. Thus, the notion that trigeminal neuralgia may present as a foregoing symptom to an early-stage tumour should not be excluded. The localisation of the tumour dictates the nature of facial symptoms. Tumours affecting the peripheral branches or the Gasserian ganglion usually give rise to sensory change and constant pain, in other words, trigeminal neuropathy [22]. Slowly growing tumours which distend rather than invade the trigeminal root are likely to cause TN. In Cheng's series, the average delay in diagnosis of the tumour was 6.3 years. Half of the patients developed sensory or motor deficits later. Trigeminal sensory deficits, bilateral involvement of the trigeminal nerve, and abnormal trigeminal reflexes are associated with an increased risk of symptomatic TN and should be considered useful in distinguishing symptomatic TN from classic trigeminal neuralgia [23]. An algorithm (Fig. 8) will assist clinicians to manage these patients without delaying the diagnosis of intracranial or extracranial tumours.

The list of differential diagnoses is long and includes a number of pathological conditions affecting the sinuses, teeth (including dental caries, root abscesses, and fractured teeth), temporomandibular joints, eyes (including glaucoma, orbital cellulitis, and trauma), nose, and the neck. Most of these are easily ruled out after the interview and brief clinical examination. Other differential diagnosis include cranial neuralgias (glossopharyngeal neuralgia, neuralgia of nervus intermedius, neuralgia of the superior laryngeal nerve, and occipital neuralgia, postherpetic neuralgia (katamnesis), reader's syndrome (lack of second trigeminal division involvement, lack of sympathetic paresis), trigeminal neuroma, ophthalmoplegic migraine, temporal arteritis, Costen's syndrome, Tolosa–Hunt syndrome (idiopathic inflammation in or around the cavernous sinus); trigeminal autonomic cephalgias (such as cluster headache and paroxysmal hemicrania)) [7, 21].

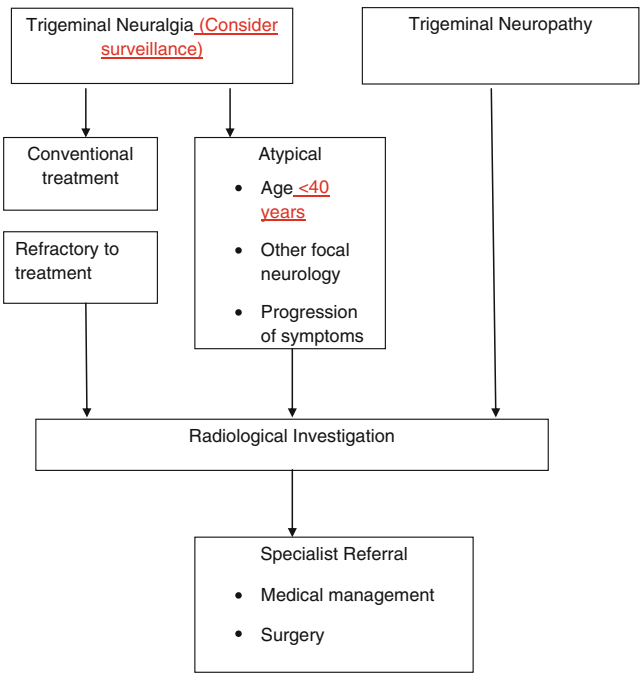

Fig. 8 Algorithm for management of trigeminal neuralgia or neuropathy

No specific tests exist for the diagnosis of TN. This should not excuse omitting the clinical examination, including assessment of cranial nerve function, given the frequency of MS and tumours found in this population. Definite facial sensory loss or other cranial nerve dysfunction, if it cannot be explained by a previously known injury to the nerve, should prompt cerebral imaging [7].

Clinical findings, in particular presence of focal neurological signs, may permit localisation of the lesion, but imaging is required to accurately localise and visualise the lesion. Radiological features may itself be diagnostic or may be used to plan stereotactic or surgical approaches. Even in typical TN, imaging studies may well be of use. In patients with TN, routine head imaging identifies structural causes in up to 15% of patients and may be considered useful [23].

In this series of cases, supplementing imaging modalities with tissue biopsies have assisted in attaining a prompt diagnosis. This is especially relevant in extracranial tumours that are more readily accessible to examination and tissue sampling. Many authors have indicated MRI scanning as an appropriate modality for ruling out demyelinating disorders and tumours [4, 7, 21]. A recent evidence-based review concluded that there is insufficient evidence to support or deny the usefulness of MRI to identify neurovascular compression [23]. Notwithstanding this, a MRI scan would be the most rational first line investigation in any patient presenting with trigeminal neuropathy as the purpose of this investigation is not only to identify vascular pathology, but also to detect other primary tumours or pathology.

Conclusion

There are several clinical features that are characteristic of trigeminal neuralgia, but these may however be misleading and masquerade the true diagnoses. This demonstrates the need for seeking the symptomatic–organic aetiology in every case not only by examination of the intracranial

trigeminal tract but also the peripheral extracranial distribution to make the correct diagnosis at the earliest possible time. Clinical findings, in particular presence of focal neurological signs, may permit localisation of the lesion, but imaging is required to potentially visualise, and accurately localise the lesion.

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Consent Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of interest statement None.

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