Case report

Arteriovenous malformation: a rare cause of trigeminal neuralgia identified by magnetic resonance imaging with constructive interference in steady state sequences

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The case

A 64-year-old female presented with a 4-year history of right-sided facial pain. She described an ‘electric shock-type’ pain starting on the right side of her chin that radiated along the lower lip, upper lip and into the lower cheek, in the mandibular division of the trigeminal nerve. The pain was triggered by speaking, eating, drinking, brushing her teeth or touching her chin. These attacks occurred daily and lasted for 3 months before resolving spontaneously, consistent with classical trigeminal neuralgia (TN). Systemic and neurological examinations were normal. In particular, trigeminal sensation was intact bilaterally and trigeminal reflexes were normal. A diagnosis of right-sided, mandibular division TN was made.

She had four similar episodes of pain over the following 4 years, each lasting ~3 months, with each episode becoming more severe. She did not want any analgesic medication for the first two episodes. However, during the most recent episode, carbamazepine (300 mg twice a day) and, subsequently, adjunctive amitriptyline (10 mg at night) were prescribed, although these failed to control the symptoms. Therefore the patient underwent investigation to assess for a structural cause amenable to neurosurgical intervention.

The patient underwent a cranial MRI with constructive interference in steady state (CISS) sequences, which demonstrated several small blood vessels surrounding the right trigeminal nerve and a very large mass, thought likely to be a venous aneurysm, lying just behind the root entry zone into the trigeminal nerve (Figure 1). A contrast-enhanced computed tomography angiogram (CTA) confirmed the previous MRI findings and was highly suggestive of an underlying arteriovenous malformation (AVM) (Figure 2). For the pre-surgical work-up a digital subtraction angiogram was organized. This demonstrated a definite AVM during contrast injection in the right vertebral artery with rapid filling of the straight sinus during the early arterial phase (Figure 3a and b).

Discussion

TN is defined by the International Headache Society (IHS) as a ‘unilateral disorder characterised by brief electric shock-like pains, abrupt in onset and termination, and limited to the distribution of one or more divisions of the trigeminal nerve’.1–3 TN is the most common neuralgia with an annual incidence of 4 to 5/100 000.1

A distinction is made between classical TN and symptomatic TN. Classical TN is defined as cases in which there is spontaneous remission and relapse of symptoms and there is no motor or sensory deficit on examination. Classical TN includes cases that are idiopathic and those secondary to vascular compression. Vascular compression, usually by the superior cerebellar artery, is the most common cause...
of classical TN and accounts for between 80% and 90% of cases.\(^4\) Symptomatic TN, due to a structural lesion other than vascular contact, presents with pain indistinguishable from classical TN in character, but the pain does not spontaneously remit and is typically associated with motor or sensory signs.\(^3\) Structural lesions identified in symptomatic TN include multiple sclerosis plaques and cerebellopontine angle tumours.\(^2\) However, classical and symptomatic TN cannot be reliably distinguished based on clinical features alone.\(^5\) Younger age of onset, abnormal trigeminal nerve evoked potentials, trigeminal sensory deficits and bilateral trigeminal nerve involvement are probably associated with an increased risk of symptomatic TN but their absence does not exclude symptomatic TN.\(^5\) Abnormal trigeminal reflexes are probably the most clinically useful with a high specificity (94%) and sensitivity (87%) for distinguishing between symptomatic and classical TN.\(^5\)

The American Academy of Neurology and European Federation of Neurological Societies (AAN–EFNS) have recently published guidelines on the management of TN.\(^2\) While medical therapy with carbamazepine or oxcarbazepine is advocated as first line therapy, the guidance recommends that early surgical intervention be considered in refractory cases. Microvascular decompression provides the longest duration of pain freedom.\(^6\) For patients in whom major surgery is not possible percutaneous procedures on the Gasserian ganglion and gamma knife surgery are effective but the recurrence of neuralgia is higher.\(^2\)

We present a case of TN not due to neuronal compression by the superior cerebellar artery, but rather compression due to an AVM. This is a rare cause of TN, accounting for \(\sim 0.22–1.8\)% of cases, and reports in the literature are limited.\(^7\)–\(^17\) One case series of 375 patients with a brain AVM between 1985 and 2004, only identified five patients presenting with TN.\(^7\) Of the five patients, four were male and one was female with age ranges between 38 and 68 years. Symptom duration varied between 4 months and 6 years. All five patients had failed conventional medical therapy with two patients also having failed percutaneous operative procedures. These observations are in accordance with the individual case reports where males were more commonly affected than females and patient ages varied between 40 and 61 years.\(^8\)–\(^13\) The duration of symptoms for patients in the case reports ranged from 6 months to 11 years with the majority of patients having failed medical therapy prior to referral for neurosurgical intervention.\(^8\)–\(^13\) Previous case
reports of patients with TN due to an AVM have not described the classical TN clinical features of spontaneous relapses and remissions, as was identified in our case.

The AAN-EFNS recommend that patients who fail medical therapy be referred for early surgical intervention and those considered suitable for surgery undergo high resolution MRI. Seven studies, selected by the AAN-EFNS, investigating the diagnostic accuracy of MRI for identifying abnormal vascular contact in classic TN demonstrated varying results. Sensitivities ranged from between 52% and 100% while specificities varied between 29% and 93%. There was significant methodological heterogeneity between these studies, including the use of different scanning protocols, which may explain some of the variation in the results. These studies demonstrate that MRI cannot recognize all cases of vascular compression in patients with TN and more importantly that not all neurovascular contact results in TN. Only one study included magnetic resonance angiography (MRA) as part of its neurosurgical work up, and reported a sensitivity of 88% and specificity of 29% for recognition of abnormal vascular contact in TN, but AVMs were not assessed specifically. Digital subtraction angiogram remains the gold standard for assessment of neck and cerebral blood vessels. However, the technique is invasive and associated with a small but significant risk of permanent neurological impairment in the order of 0.09–0.5%

The AVM in our patient was identified using MRI with CISS sequences. Two reports have demonstrated MRI with CISS to be the superior imaging modality for evaluation of TN. Yamakami et al. compared MRI-CISS against MRI with three-dimensional fast inflow with steady-state precession (FISP) in patients with TN and hemifacial spasm. They found that MRI-CISS imaging was superior to FISP with higher resolution and excellent contrast demonstrating precisely the anatomic detail and abnormal neurovascular relationships responsible for TN. A study in Japan evaluated MRI-CISS and MRA for detection of neurovascular compression and assessed the relationship between clinical symptoms and the site of trigeminal nerve compression. MRI-CISS was able to identify both arterial and venous compression of the trigeminal nerve whereas MRA could not detect the vein as the cause of compression in TN. The authors demonstrated that the findings from MRI-CISS imaging were identical to those encountered upon surgery and that a close relationship was found between the region of neuralgic manifestation, where corresponding trigeminal branch fibres are distributed, and the site of vascular compression in the trigeminal nerve.

As TN secondary to AVM occurs infrequently consensus regarding best treatment has not been reached. Treatment options include embolization of the AVM to reduce its size and blood flow using endovascular coils or ethylene-vinyl alcohol copolymer injections before resection of the AVM. Both case reports of embolization of the AVM demonstrated resolution of facial pain. Garcia-Pastor et al. report the successful use of microvascular decompression in four patients with the complete disappearance of the neuralgia. However, one patient suffered a haemorrhage from the AVM during the microvascular decompression and the procedure could not be completed.

Although larger case series or cohort studies are needed, this case demonstrates that, even in patients with symptoms and signs of classical TN, MRI-CISS imaging may be a pragmatic non-invasive screening technique for the diagnosis of AVM-induced trigeminal neuralgia.
tool by which to identify vascular lesions such as an AVM.

Conflict of interest: None declared.

References


