

CONTINUING EDUCATION PROGRAM: FOCUS...

Trigeminal neuralgia



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KEYWORDS

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Abstract Two different clinical entities, essential or secondary neuralgia, are associated with different pathologies. The pathways of CN V comprise the cervical spine, the brainstem, the root of the nerve and the three peripheral branches: V1, V2 and V3. The lesions responsible for neuralgia are neoplastic, vascular, inflammatory, malformative or post-traumatic. The examination protocol should explore the set of CN V pathways. Neurovascular compression is the main cause of essential neuralgia. It is investigated by T2-weighted inframillimetric volume. Two conditions are necessary to diagnose a neurovascular compression: localised on the root entry zone [(REZ), 2–6 mm from the emergence of the pons] and perpendicularly. In the absence of neurovascular compression, thin slices and a gadolinium injection are necessary.

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Trigeminal neuralgias can be extremely severe facial pain of a highly debilitating nature. Imaging is indicated to identify any curable lesion. Trigeminal neuralgia can be caused by multiple lesions on the pathways of the fifth cranial nerve pair: the sensory nuclei of CN V, the sensory root and its branches, up to the skin.

As a result of this, the assessment of trigeminal neuralgia requires a good understanding of anatomy and possible aetiological ranges.

Objectives

The objectives are as follows:

- clinical understanding and physiopathology of the two types of trigeminal neuralgia;
- knowledge of the normal anatomy of the trigeminal nerve;
- knowledge of the principle lesions responsible for trigeminal neuralgia;
- knowledge of how to protocol an MRI examination for trigeminal neuralgia;
- knowledge of the principle therapeutic option.

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Anatomy

The trigeminal nerve is the fifth cranial pair. It is a sensory and motor nerve, consisting of a large sensory root and a small motor root.

The sensory root transmits sensory information from the unilateral hemiside and is divided into three branches which correspond to three different skin areas (dermatomes): V1, V2 and V3 (Fig. 1).

The motor root innervates the unilateral masticator muscles. This root cannot be distinguished from the sensory root in MRI. It has a common pathway with the common trunk of the nerve and root of CN V3 over the whole pathway.

Trigeminal neuralgia can be caused by a lesion of the sensory nuclei of CN V and the sensory root and its branches up to the skin.

Exploration of the pathways of CN V, therefore, involves analysis of the posterior fossa and the facial bones [1].

The nuclei of CN V have broad distribution over the brainstem and the upper cervical spine (Fig. 2). This explains why cervical medullar, bulbar, pons and mesencephalic lesions can be responsible for CN V neuralgia.

The root of CN V emerges from the pons and follows a pathway within the base cisterns to Meckel's cave (Fig. 3).

This cave corresponds to a fold of the dura mater, containing cerebrospinal fluid. Within Meckel's cave, we find Gasser's ganglion, which corresponds to the division of the root of CN V into three branches (Fig. 4).

Meckel's ganglion is not associated with significant enlargement of the root ending.

Branch V3 and the motor root present a vertical pathway, descending towards the masticator space through the foramen ovale (Fig. 5). This descending pathway takes place at the junction between the termination of Meckel's cave and the entry point into the cavernous sinus. Therefore, CN V3 does not cross the cavernous sinus.

Branches V1 and V2 have an anterior horizontal pathway on the lateral side of the cavernous sinus.

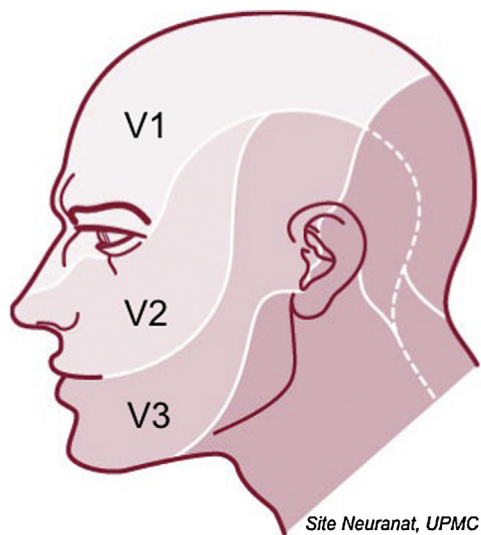


Figure 1. Representation of dermatomes corresponding to the three CN V branches: V1, V2 and V3.



Figure 2. Representation of CN V nuclei: sensory nuclei and their afference are represented in grey, the motor nucleus and its efference are represented in white.

At the end of the cavernous sinus, branch V2 crosses three structures: the foramen rotundum (Fig. 6), the pterygopalatine fossa (Fig. 7) and then, the infraorbital canal (Fig. 8) up to the skin.

Branch V1 crosses the superior orbital fissure and presents a pathway within the orbit (Fig. 9).

The distal ends of the branches correspond to the dermatomes in Fig. 1.

Clinical presentation

Two different clinical pictures emerge which correspond to different aetiological ranges.

Essential neuralgia

Essential neuralgia has a characteristic clinical presentation. Patients present with intense paroxysmal pain of the electrical discharge type ("painful flashes"), which lasts about a second and which occurs in repeated bursts for several minutes. This pain is always unilateral and typically limited to one or two branches: usually V2 and V3.

In some patients, these painful bursts can be triggered simply by cutaneous contact with a trigger zone or during movement. Where this trigger zone exists, there is a refractory period at the end of an episode of pain during which the subject can touch the trigger zone without this causing pain.

Neurological examination of these patients is usually normal.

Pain is alleviated by inhibitors of the sodium channels.

Neurovascular compression is the principal cause of essential neuralgia.

Other causes are possible, such as Chiari's malformation, multiple sclerosis or a lesion of the cerebellopontine angle stretching the root.

Some patients have a normal MRI scan.

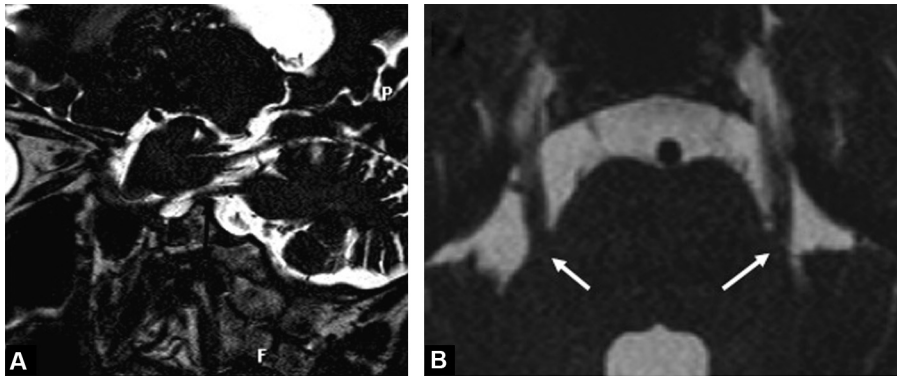


Figure 3. Visualisation of the roots of CN V within the base cisterns, after their emergence from the pons on a T2-weighted inframillimetric sequence in the sagittal (A) and axial (B) planes.

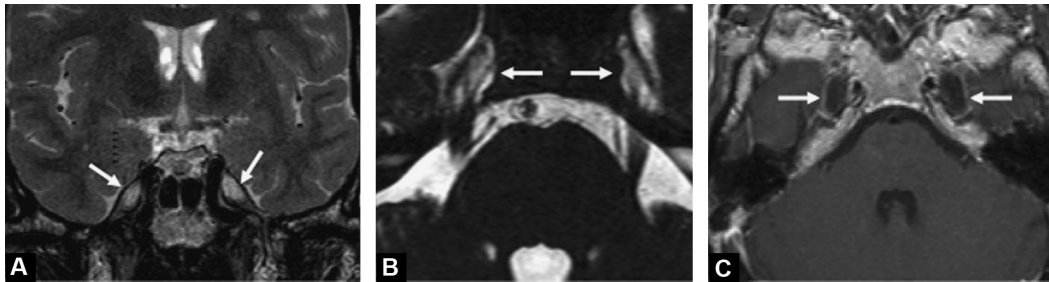


Figure 4. Visualisation of Meckel's cave (trigeminal fossa) bilaterally in T2 coronal (A), T2-weighted axial (B) and T1 after injection (C). This cavity corresponds to a fold in the dura mater containing the CSF and Gasser's ganglion. The division branches of CN V are visible on T2-weighted sequences (B).

Secondary neuralgia

The clinical picture differs from essential neuralgia. The pain is usually dull, continuous, with painful paroxysms lasting 3–4 h. The disorder can affect the three dermatomes. There is no trigger zone.

The neurological examination may be normal: associated hypoesthesia, impairment of other cranial nerves or of the central nervous system.

There are many aetiologies and any pathology on the pathway of CN V that can be responsible for secondary neuralgia.

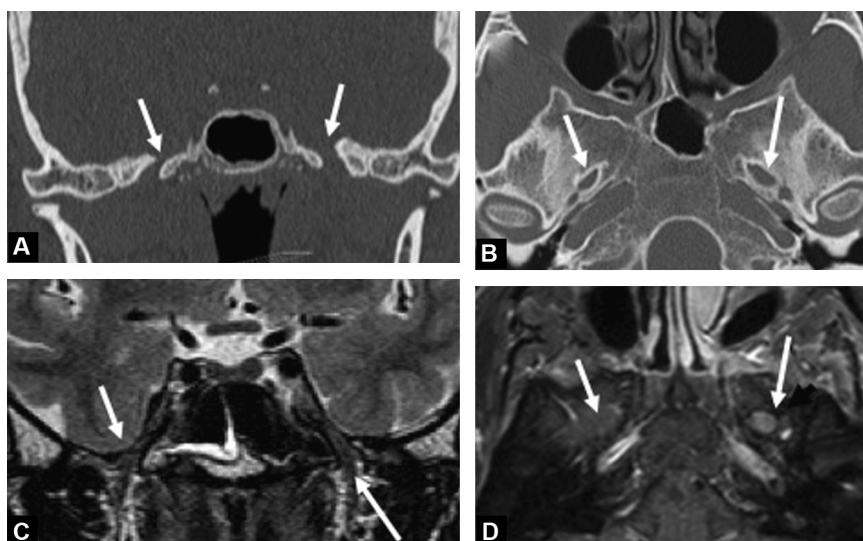


Figure 5. Visualisation of the oval foramen (arrows) through which CN V3 passes. Scan image in the coronal (A) and axial (B) sections and views corresponding to T2 coronal (C) and T1 axial after injection with fat saturation (D). On image D, the left oval foramen is enhanced abnormally as a result of tumour infiltration (perineural extension) whereas the healthy contralateral foramen ovale is not visible after injection.

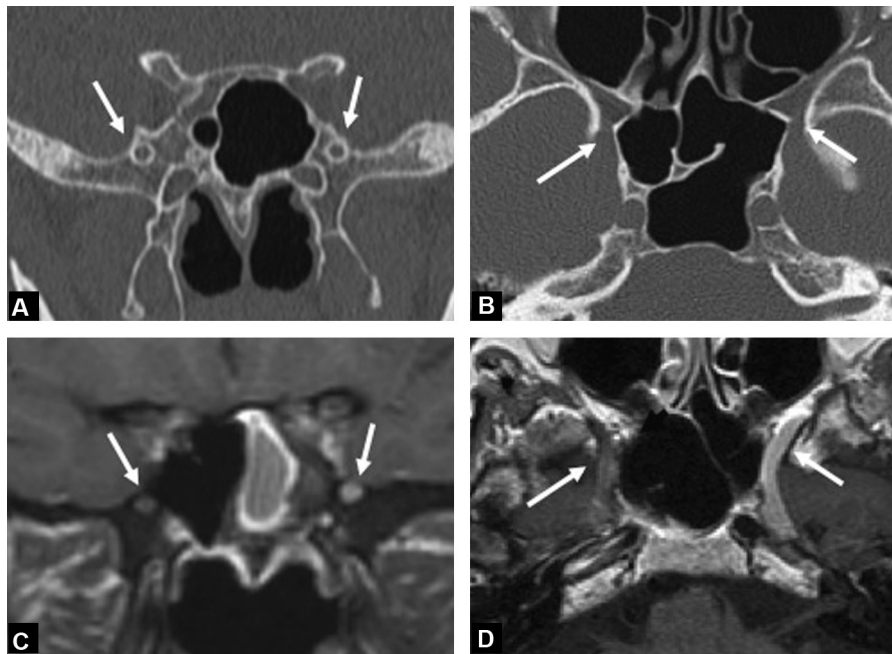


Figure 6. Visualisation of the foramen rotundum through which CN V2 passes (arrows). Scan image in the coronal (A) and axial (B) sections and views corresponding to T2 coronal (C) and T1 axial after injection with fat saturation (D). On images C and D, the left rotundum foramen is enhanced abnormally as a result of tumour infiltration (perineural extension) whereas the healthy controlateral foramen rotundum is not visible after injection.

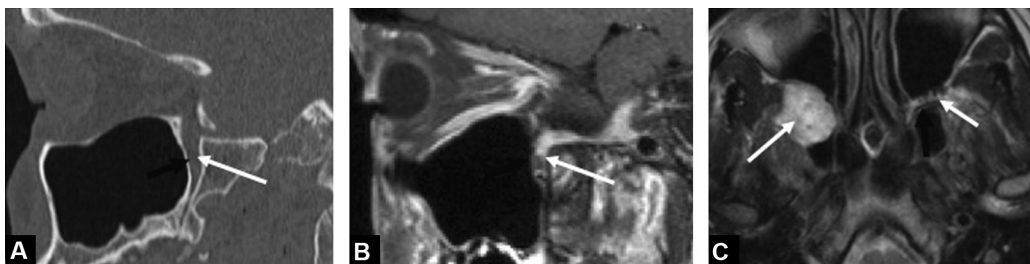


Figure 7. Visualisation of the pterygopalatine fossa which CN V2 passes (arrows). Scan image in the sagittal plane (A), T1 sagittal view after injection with fat saturation (B) and T2 axial view (C). On image B, the foramen rotundum is enhanced abnormally as a result of tumour infiltration (perineural extension). On image C, a regular hyper T2 mass corresponding to a Schwannoma is present in the right pterygopalatine fossa.

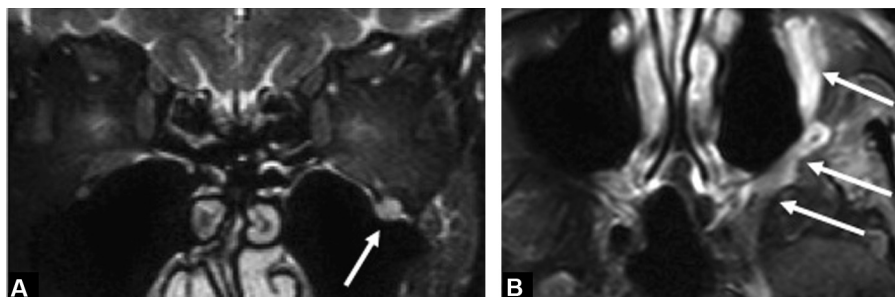


Figure 8. Visualisation of the infraorbital canal through which V2 passes (arrows). T1 coronal (A) and axial (B) views after injection with fat saturation. The left infraorbital canal is abnormally enhanced due to an infiltration (sarcoidosis) whereas the healthy controlateral canal is not enhanced after injection. On image B, this infiltration is visualised over the pathway of CN V2: continuously with the cavernous sinus, round foramen and pterygopalatine fossa (arrows).

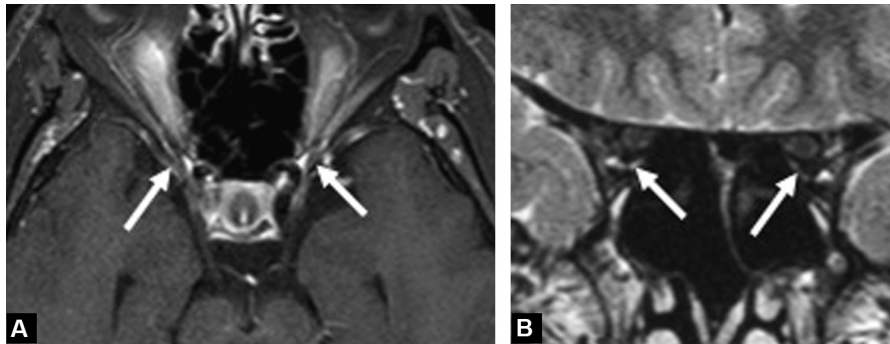


Figure 9. Visualisation of the superior orbital fissure crossed by CN V1 (arrows). T1 axial view after injection with fat saturation (A) and T2 coronal (B).

Investigation of trigeminal neuralgia

MRI investigation, and therefore the protocol, may vary as a function of the suspected pathology and abnormalities observed during the examination.

An initial protocol can be suggested:

- T1 sagittal (Fig. 10): this makes it possible to locate the root of CN V, usually in the first lateral slice to the brainstem, at the level of the pons. Thin slice acquisitions will be positioned along the axis of the nerve. Also, this sequence explores the cervico-occipital joint;
- T2 axial: explores the brainstem, cavernous sinus, facial bones;
- T2-weighted axial centred on CN V;
- at least one injected section of thin slices passing through CN V, depending on the results, other sequences may be carried out;
- in the case of neurovascular compression, carry out 3D TOF vascular acquisitions to determine the arterial or venous source of the conflict.

Aetiologies

Essential neuralgia

Several causes of essential neuralgia can be observed:

- neurovascular compression, which represents the principle aetiology responsible for essential neuralgia (Fig. 11);



Figure 11. Left neurovascular compression (arrow). Perpendicular vessel leading to pressure on the nerve on the zone corresponding to REZ (2 mm from the emergence of the brainstem).

- multiple sclerosis (Fig. 12);
- a lesion developing in the cisterns stretching the root of CN V (Fig. 13);
- Chiari's malformation.

Finally, neuralgia can be purely essential, in other words, without a causal lesion found during investigations.

Neurovascular compression

The physiopathology of neurovascular compression is poorly understood. Repeated microtraumas linked to vascular pulsation may induce a demyelination zone with aberrant

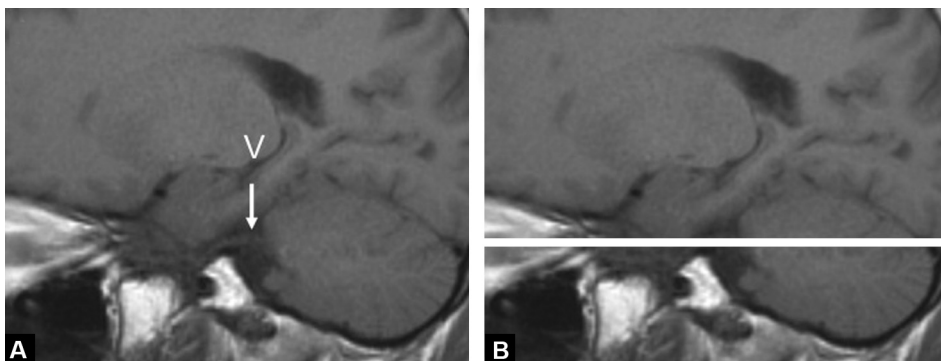


Figure 10. Visualisation of the root of CN V on the first lateral cut at the brainstem with T1 sagittal acquisition (A, arrow) and representation of the axis of the plane of acquisition chosen to position fine cut sequences (B).

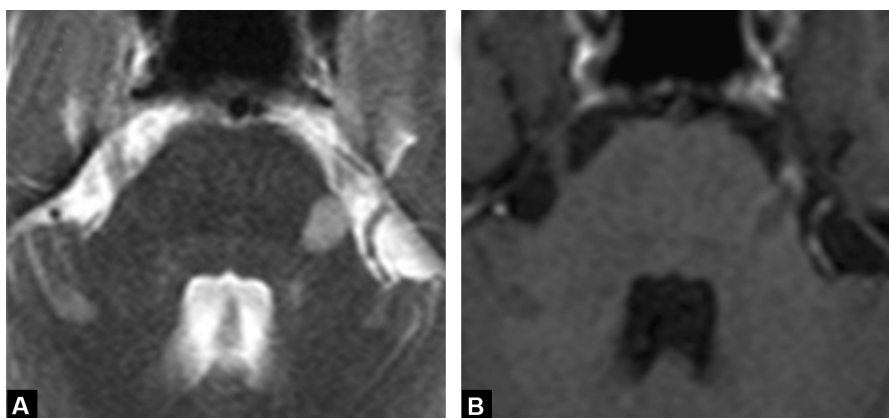


Figure 12. Multiple sclerosis lesion responsible for left CN V neuralgia of the essential type in T2 axial (A) and T1 after injection (B). Interestingly, enhancement of the first millimetres of the root of CN V is observed (central myelin zone ahead of REZ).

remyelination and the creation of neoreceptors, which can generate ectopic influxes.

This conflict can only take place in a precise area of the nerve, which corresponds to a fragile area of the nerve called root entry zone (REZ) or transition zone. It corresponds to the transition zone between central myelin (oligodendroglia) and peripheral myelin (Schwann cells). This transition does not take place at the emerging point of the nerve but in the nerve root at a varying distance depending on the nerve. For the trigeminal nerve, REZ is 2–6 mm from emergence of the brainstem.

To confirm diagnosis of neurovascular compression, two conditions are necessary:

- contact of a vessel (artery or vein) with REZ;
- perpendicular to the axis of the nerve.

At best, this contact is associated with a mass effect (pressure) on the nerve's pathway (Fig. 11) [2].

Secondary neuralgia

All diseases of the CN V pathway may be responsible for secondary neuralgia. Good understanding of anatomy is therefore essential to explore the set of zones where this abnormality can occur.

Understanding of the dermatome(s) involved is often an essential aid to diagnosis.

We are able to observe the following lesions as a function of topography [3–5].

Lesion of the nuclei of V

There are many pathological conditions that involve the brainstem and the cervical spine. The most commonly observed lesions are ischaemia, haemorrhage (cavernoma), multiple sclerosis, tumour and syringomyelic cavitation (Fig. 14).

Lesions of the root within the cisterns and Meckel's cave

The appearance of lesions can be non-specific (Fig. 15). In fact, we can see continuous staining from the root in pathologies as varied as infectious meningitis (CMV, tuberculosis), carcinomatous meningitis, lymphoma or neurosarcoidosis. In these situations, diagnosis is based on associated localisations or a general exploratory assessment.

In this topography, impairment of the root may be either benign in origin (sarcoidosis, Schwannoma) or malignant (metastases, perineural extension, lymphoma or carcinomatous meningitis). The root can also be the site of extrinsic

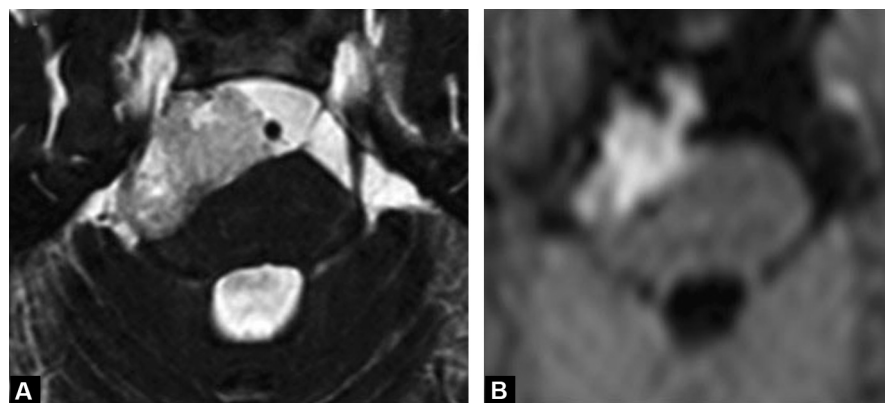


Figure 13. Epidermoid cyst revealed by essential neuralgia of the right CN V. The root of right CN V is stretched by the cyst on the T2 inframillimetric image (A). Axial diffusion (B) confirms diagnosis of an epidermoid cyst.

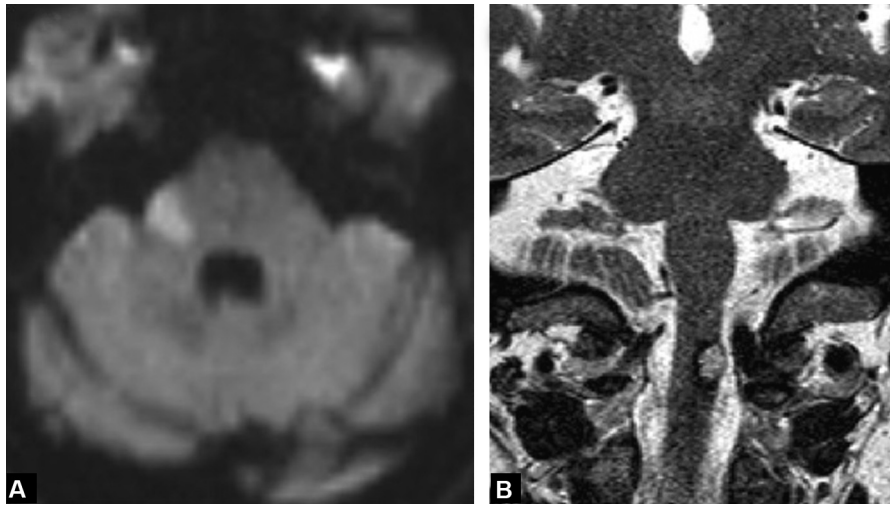


Figure 14. Examples of secondary neuralgia caused by lesions of the CN V nuclei. Ischaemic stroke in axial diffusion (A) and left cervical medullary cavernome in T2 coronal (B).

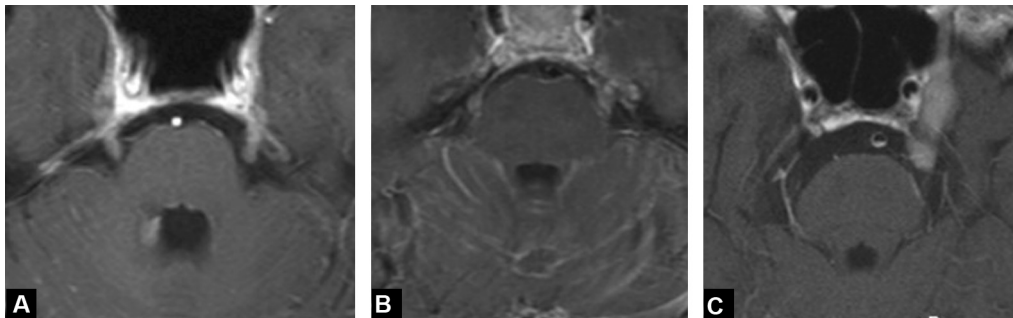


Figure 15. Examples of secondary neuralgia associated with non-specific contrast uptake of the CN V roots in injected T1 axial: lymphoma (A), carcinomatous meningitis (B) and discontinuous perineural extension of a cystic adenoid carcinoma (C).

compression (epidermoid cyst, Schwannoma of CN VIII) (Fig. 16).

Lesions of the cavernous sinus

Lesions of the cavernous sinus can be primary, such as a meningioma, Schwannoma or an aneurysm. The localisation of sarcoidosis, lymphoma or metastases is often very non-specific (Fig. 17).

Finally, the lesion may be found to be an extension into the cavernous sinus of a lesion on the base of the skull (chondromas, chordomas, metastases) or a lesion of an ENT lesion by perineural extension that is either continuous (by

proximity) (Fig. 18A) or discontinuous (cystic adenoid carcinomas) (Fig. 18B).

Distal branches lesions

Schwannomas are observed (Fig. 7C).

The other impairments are usually non-specific: continuous contrast medium uptake, T2 hyposignal: sarcoidosis (Figs. 6C and D, 8A and B) or perineural extension of an ENT cancer (Fig. 5D, 7B, 19).

In the case of CN V3, as a result of the common pathway of the sensory root with the motor root, unilateral amyotrophy of the masticator muscles may be observed (Fig. 19B).



Figure 16. Examples of secondary neuralgia caused by a lesion on the root of CN V (A, B) and Meckel's cave (C): compression by a Schwannoma of the right CN VIII (A), Schwannoma of the left CN V (B) and infiltration of the left Meckel's cave by sarcoidosis (C).



Figure 17. Examples of left secondary neuralgia caused by lesions of the cavernous sinus: carotid aneurysm (A), meningioma (B), sarcoidosis (C).

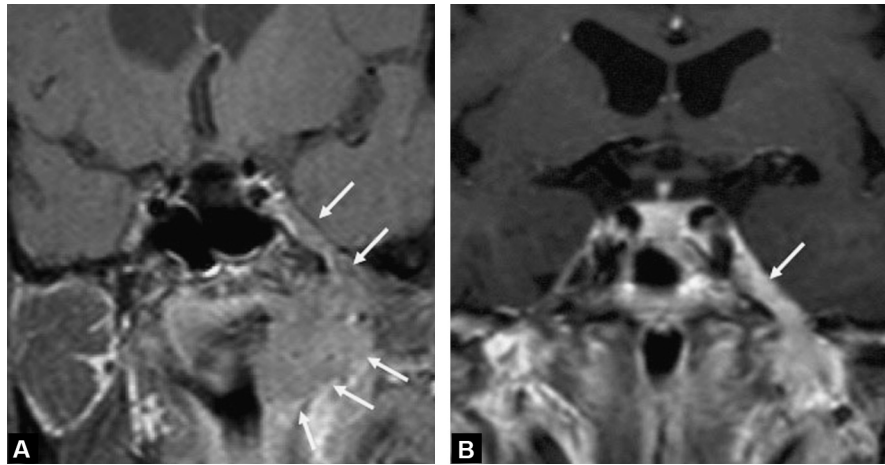


Figure 18. Examples of perineural infiltration of the cavernous sinus and oval foramen. Continuous perineural extension (A) of a UCNT in the left cavernous sinus by the oval foramen (arrows). Discontinuous perineural extension (B) of a cystic adenoid carcinoma.

This element reveals the presence of a lesion (sometimes millimetric) on the pathway of CN V3.

As a result of the broad range of possible pathologies, analysis of various areas of CN V, the cervical spine and the facial region is essential.

The imaging protocol is often modified in the course of examination in order to be able to carry out modified complementary sequences (fat saturation, thin slices, etc.).

Treatment

Medical

Initially, treatment is medical and often effective.

The most commonly used molecules are carbamazepine, oxcarbazepine, gabapentin or lamotrigine.

Surgical

In the case of a failure of medical treatment, two main techniques are suggested: thermocoagulation of Gasser’s ganglion and neurovascular decompression.

Rhizolysis

The most common technique is thermocoagulation of Gasser’s ganglion by transcutaneous route. This technique is effective for a dozen years and can be repeated two or three times in a patient’s lifetime. Following this, there is a risk of denervation pain, which can be equally debilitating.

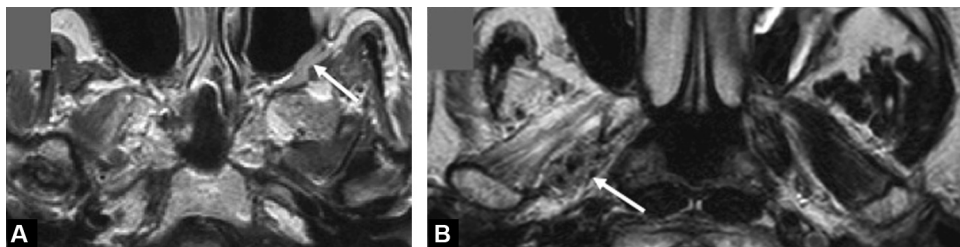


Figure 19. Examples of secondary neuralgia caused by small size discontinuous perineural extensions (arrows). Impairment of left CN V2 in the pterygopalatine fossa (A). Impairment of the right CN V3 in the masticator space (B): amyotrophy of the unilateral masticators indicates the presence of a lesion on CN V3.

Neurovascular decompression

In the case of neurovascular compression, a surgical approach can be carried out to eliminate the compression, and usually involves inserting a material (Teflon plate) between the vessel and the root.

Other techniques

Some neuralgia conditions can be treated by radiosurgery. Sometimes alcohol therapy of the peripheral branches is indicated.

Conclusion

The range of pathologies responsible for trigeminal neuralgia is vast and the possible topographical areas of impairment are extensive ranging from the cervical spine to the face and skull base.

MRI investigation is based on an initial imaging protocol that can be completed by examination if lesions are detected.

In this context, clinical understanding of the patient prior to the MRI scan is often a precious aid in directing the examination (essential or secondary neuralgia, affected dermatomes).

In the case of essential neuralgia, the most commonly observed lesion is neurovascular compression. Diagnosis is based on T2-weighted inframillimetric acquisition by visualising a vessel perpendicular to the REZ (at 2–6 mm from the emergence of the brainstem). 3D TOF then makes it possible to determine the arterial or venous origin of the compression.

In the case of secondary neuralgia, a good understanding of the anatomy of the CN V pathways is necessary.

TAKE-HOME MESSAGES

- The range of pathologies responsible for trigeminal neuralgia is vast: neoplastic, vascular, inflammatory, malformative or post-traumatic.
- Two different clinical entities: essential or secondary neuralgia are associated with different pathologies.

- The possible topographical area affected is extensive from the cervical spine to the facial region.
- MRI investigation is based on an initial imaging protocol that can be completed by examination if lesions are detected.
- In this context, clinical understanding of the patient prior to the MRI scan is often a precious aid in directing the examination (essential or secondary neuralgia, affected dermatomes).
- In the case of essential neuralgia, the most commonly observed lesion is neurovascular compression. Diagnosis is based on T2 inframillimetric acquisition by visualising a vessel perpendicular to the REZ (at 2–6 mm from the emergence of the brainstem). 3D TOF then makes it possible to determine the arterial or venous origin of the compression.
- In the case of secondary neuralgia, good understanding of the anatomy of the CN V pathways is necessary.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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