



Typical Trigeminal Neuralgia by an Atypical Compression: Case Report and Review of the Literature

Atipik Kompresyonun Neden Olduğu Tipik Trigeminal Nevralji: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

AIM: Trigeminal neuralgia is a disabling form of facial pain that causes a considerable discomfort and a marked reduction in the quality of life. Although neurovascular compression is often associated with trigeminal neuralgia, other intracranial pathologies can result in compression along the nerve.

CASE DESCRIPTION: The authors report a case of a patient suffering from a typical left trigeminal neuralgia affecting both the second and the third branches. Neuroradiological examinations showed the presence of an arachnoid cyst located in the left temporal lobe, contiguous with the Meckel's cave, eroding the greater wing of the sphenoid bone. Following a meticulous decision-making process the patient underwent conservative treatment with success. A properly modulation of the drug dosage allowed pain relief to be achieved.

CONCLUSION: This is a rare case reporting a typical trigeminal neuralgia by an arachnoid cyst of the Meckel's cave mimicking a neurovascular compression syndrome. Our findings, behind the review of the current literature, point out the importance of such a pathological condition when dealing with patients affected by cranial nerve dysfunction syndrome.

KEYWORDS: Arachnoid cyst, Trigeminal neuralgia, Conservative treatment

ÖZ

AMAÇ: Trigeminal nevralsi önemli rahatsızlığa ve yaşam kalitesinde belirgin azalmaya yol açan, maluliyete neden olan bir fasyal ağrı tipidir. Nörovasküler kompresyon trigeminal nevralsi ile sıklıkla ilişkili olsa da başka intrakraniyal patolojiler de sinir kompresyonuna yol açabilir.

OLGU TANIMI: Yazarlar hem ikinci hem üçüncü dalları etkileyen tipik sol trigeminal nevralsisi olan bir hastayı bildirmektedir. Nöroradyolojik incelemeler sol temporal lobda Meckel boşluğuyla devam eden ve sfenoid kemiğin büyük kanadında erozyona yol açan araknoid kist varlığını gösterdi. Kapsamlı bir karar verme sürecinden sonra hastaya başarıyla konservatif tedavi uygulandı. İlaç dozajının uygun şekilde modülasyonu ağrının geçmesini sağladı.

SONUÇ: Bu olgu, nadir bir nörovasküler kompresyon sendromuna benzeyecek şekilde Meckel boşluğunun araknoid kisti nedeniyle tipik bir trigeminal nevralsi olgusudur. Bulgularımız mevcut literatürle değerlendirildiğinde kraniyal sinir disfonksiyonu sendromu olan hastalarda böyle bir patolojik durumun önemini ortaya koymaktadır.

ANAHTAR SÖZCÜKLER: Araknoid kist, Trigeminal nevralsi, Konservatif tedavi

INTRODUCTION

Typical trigeminal neuralgia (TN) is defined as paroxysmal and/or recurrent attack of pain lasting from fraction of second to few minutes, involving one or more divisions of fifth cranial nerve.

The annual incidence is approximately 4.5 per 100 000 in the general population, with a female/male ratio of 3/2 (12).

Clinical features of TN have been extensively described in worldwide literature. Pain is typically described as unbearable,

lancinating, stabbing or superficial, causing a discomfort to the patient. Painful attacks are often primed by light sensory stimulation (touching, drinking, eating, shaving, applying make-up) of the so called trigger zones.

According to the International Classification of Headache Disorders, TN in its etiology is divided into classic form, caused by vascular compression of trigeminal nerve root, and symptomatic one, caused by other factors as tumors, vascular disorders, demyelination in multiple sclerosis (1). To date, only two cases of TN, related to the presence of an arachnoid

cyst, have been described (14,15). Our report adds a new case with unique features and points out the importance of such a pathological condition when dealing with patients affected by cranial nerve dysfunction syndrome.

The patient has consented regarding publication of the medical data.

CASES REPORT

Clinical Presentation

A 40-year-old white male presented with two-years-long history of left facial pain on V2 and V3 branches of the V cranial nerve. Pain was described as stabbing, chronic with intermittent poussées and not related to seasonality. Stimulation of trigger zones by chewing, light touching or shaving, caused the onset of pain. Non steroid anti-inflammatory drugs and, subsequently, steroids were not effective in pain relief. Half-

left face dysesthesia and hyperpathia accompanied the painful attacks. Over the time, symptoms worsened thus forcing the patient in assuming a semi-liquid diet with a deterioration of the quality of life.

Magnetic Resonance Imaging (MRI), with specific angiographic sequences, did not disclose the presence of a neurovascular compression. T1- and T2 -weighted MRI showed a lesion nearby the left temporal lobe in the Meckel's cave region. It was isointense with cerebrospinal fluid. The lesion presented with an anterior intra-diploic extension into the left greater wing of the sphenoid bone (Figure 1A,B). These neuroradiological findings suggested that the lesion was an arachnoid cyst and that it was causing the patient's trigeminal neuralgia. Multi-slice Computer Tomography (CT) scan clearly showed a bony erosion with a defect in the floor of the middle cranial fossa (Figure 2A,B).

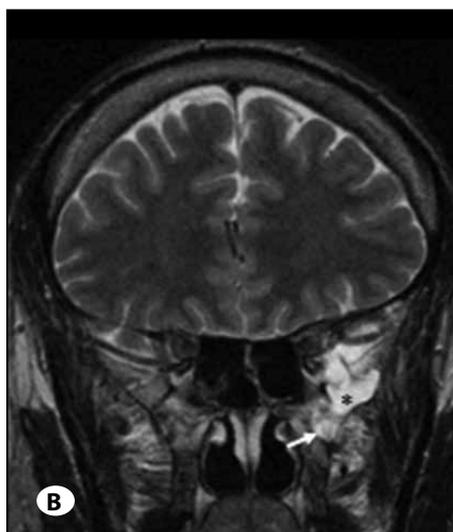
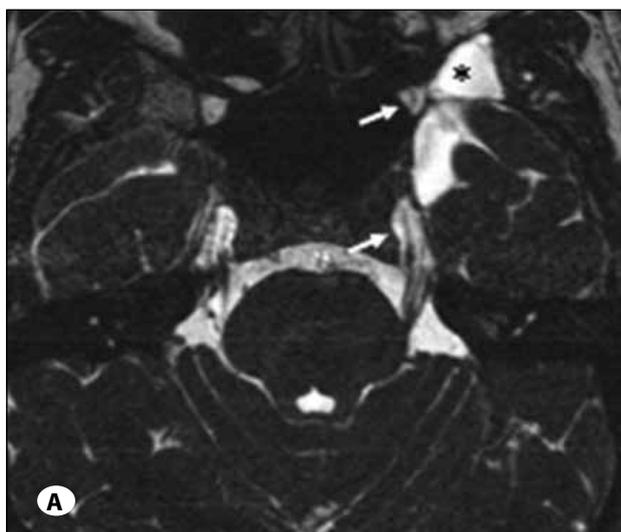


Figure 1: MRI examination: Axial CISS T2-weighted image (A) and Coronal High-Resolution FSE T2-weighted image (B). A small arachnoid cyst is present at left temporal lobe. The lesion has an anterior intradiploic extension (asterisk) at the greater wing of the sphenoid bone. Note the close relationship with the Meckel's cave and V2 root (arrows).

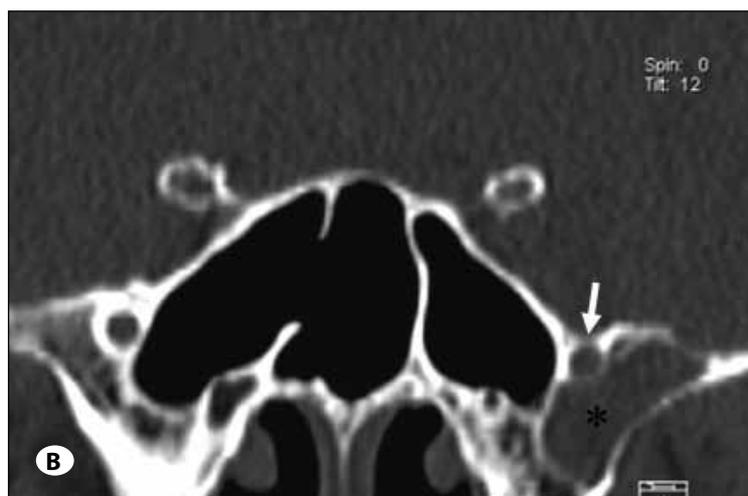


Figure 2: Multislice-CT scan: Sagittal (A) and coronal (B) reconstruction. A bony defect (arrow) is clearly depicted. It represents the communication between intracranial and extracranial component of the cyst (A). Note the close proximity between intrasosseous cyst (asterisk) and demineralised round foramen (arrow) (B).

Table I: Cases of Symptomatic Compression of the V Cranial Nerve by Arachnoid Cyst

Authors	Patient Age (yrs)	Gender	Cyst location	Treatment
Genc E et al. (8)	22	Male	Prepontine clival	Conservative
Mitsos AP et al. (13)	68	Female	Quadrigeminal	Surgery *
Ohnishi YI et al. (14)	62	Female	Quadrigeminal	Surgery **
Sugita K et al. (15)	50	Male	Middle cranial fossa	Surgery ***
Grasso G et al.	40	Male	Meckel's cave	Conservative

* Cyst fenestration;

** Neuroendoscopically-assisted cyst-cisternal shunting;

*** Cystoperitoneal shunting.

Treatment

Since the cyst was small in size and, besides the left fifth cranial nerve dysfunction, it did not cause intracranial hypertension a conservative treatment was considered as the first step in the decision-making process. Patient received medical therapy, with progressively increasing doses of carbamazepine, to obtain complete control of pain. The carbamazepine dosage was stabilized at 1200 mg per day, distributed into three separated doses. At two year follow-up the patient was free of symptoms excepting for occasional facial dysesthesia.

DISCUSSION

Several theories have been developed regarding the pathophysiological mechanisms underlying the onset of trigeminal neuralgia. The concept of vascular compression of cranial nerves in the posterior fossa has developed from several lines of evidence. Dandy first proposed the fifth cranial nerve compression, at its point of entry into the pons, by the superior cerebellar artery, as a possible cause of trigeminal neuralgia (6). Subsequent reports confirmed that patients with trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia had blood vessels in close contact with the respective cranial nerve, and that separating the blood vessel from the nerve by interposing a soft implant between them (microvascular decompression) could be curative (7,10,11).

Pathophysiological mechanisms underlying cranial nerve hyperactive dysfunction after vascular compression have been investigated extensively and partially clarified. Briefly, it has been suggested that these clinical syndromes result from pulsatile compression by arteries at the root entry/exit zone of the cranial nerve, a junctional area between central and peripheral myelin (11). Over the past several years, this concept has been widely accepted and has stimulated several studies addressed primarily at establishing precise patient selection criteria. With the advent of magnetic resonance imaging, which, using specific three-dimensional sequences, has offered a good visualization of both cranial nerves and cerebral vessels, neurovascular compression disorders have been diagnosed with increasing frequency, thus providing

additional evidence supporting microvascular decompression treatment.

In some cases, distortion of the trigeminal nerve roots by various lesions can lead to trigeminal neuralgia (3-5,9).

Our patient's trigeminal neuralgia has been caused by marked compression of the V cranial nerve at the Meckel's cave area. The literature search identified only one case report dealing with a middle cranial fossa arachnoid cyst causing trigeminal neuralgia (15) and three cases of posterior cranial fossa arachnoid cyst causing symptomatic compression of the fifth cranial nerve (8,13,14) (Table I). Arachnoid cysts have been also associated with other cranial nerves dysfunction in some cases (2).

Arachnoid cysts are congenital lesions arising from an anomalous splitting in two layers of the arachnoid membrane which constitute an area collecting the cerebrospinal fluid (CSF). The cyst can communicate with the normal CSF circulation system or, by a "valve mechanism", can grow. Conceivably, pulsatile and rhythmic variations of intracranial pressure, synchronous with the heartbeat, could be responsible for arachnoid cyst pulsation that, mimicking the mechanism of the neurovascular compression, may produce neural damage.

Our literature search identified only one case report dealing with the association of middle cranial fossa cyst and TN (15). However, based on the report, it was unclear whether the TN was with a typical or atypical clinical presentation and if the medical therapy was unsuccessful. In that case surgical treatment of cystoperitoneal shunt was performed with pain relief obtained six months later.

Our case is unique since the TN, caused by an arachnoid cyst compressing the nerve at the Meckel's cave, has been successfully managed by medical therapy. Given the peculiarity of our findings, the reported case points out the importance of the knowledge of such a pathological condition when dealing with patients affected by cranial nerve dysfunction syndrome and suggests a conservative treatment in case of drug responsiveness and non-hypertensive cyst.

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