A cluster-tic syndrome: a case report

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Abstract

Introduction: Cluster headaches and trigeminal neuralgia have typical clinical features with recurrent, attacks of severe pain in orbito-temporal area along with unilateral lacrimation and rhinorrhea (cluster headache) or within the innervation area of the roots of trigeminal nerve branches (trigeminal neuralgia). Co-occurrence of these two conditions, as it is often referred in the literature, is named cluster-tic syndrome.

Case report: A 46-year-old, female patient was presented with episodic form of cluster headaches and concurrent trigeminal neuralgia. Painful attacks had a different time distribution and matching localization. Specific treatment led to relief in both types of headaches. MRI of the brain showed aberrant superior cerebellar artery which contacts with proximal segments of right trigeminal nerve, possibly leading to neuropathic pain.

Discussion: Cluster headache has characteristic clinical presentation with intermittent, recurrent, severe pain and dysautonomic features in the orbito-temporal area, specific treatment of acute pain attacks (oxygen inhalation, subcutaneous sumatriptan injections) and specific prevention therapy (verapamil, prednisone), different from other primary headaches. Trigeminal neuralgia is presented with attacks of stabbing unilateral facial pain following the sensory distribution of one or more of the branches of trigeminal nerve. Both conditions are rare and its association is even more rare and called cluster-tic syndrome and they produce a strong and exhausting pain which requires immediate attention and treatment.

Keywords: Cluster headache, Trigeminal neuralgia, Cluster-tic syndrome.
**Introduction**

Cluster headache is characterized by unilateral, recurrent, short-lived severe pain in the orbito-temporal area along with ipsilateral autonomic symptoms (lacrimation, rhinorrhea). Trigeminal neuralgia is characterized by recurrent, unilateral attacks of sharp, stabbing pain in the sensory innervation of one or more trigeminal nerve branches [1].

The cluster-tic syndrome is characterized by the coexistence of two kinds of pain. One is strictly unilateral, usually periorcular, with evident autonomic features, and daily attacks for weeks or months (cluster). The other is characterized by paroxysms similar to electric shocks—tics (trigeminal-like) [2].

**Case report**

We present a female patient with episodic form of cluster headache in the right orbito-temporal area which gradually developed typical features of trigeminal neuralgia with the same distribution. MRI of the brain showed aberrant superior cerebellar artery which contacts proximal segments of right trigeminal nerve.

A 46-year-old female patient had three episodes of cluster headaches which lasted for a month and were repeated biannually. The pain was strong, sharp, localized in right orbito-temporal area. Usually occurring around 16:30h, lasting for 15-30 minutes and repeating at 6-8 occasions before stopping around 23:30. During the pain attacks, the patient was experiencing ipsilateral lacrimation and nasal congestion (autonomic dysfunction). The patient was restless and constantly in motion. Neurological findings were normal. CT of the head was normal.

During the attacks, we administered 100% oxygen via nasal catheter (8-10 L/min for 15-20 minutes) and subcutaneous sumatriptan (6 mg). For preventive treatment verapamil and prednison were introduced from the start. Treatment with verapamil lasted for two additional weeks (80 mg tid) while prednisone, after it was started, at initial dose of 60 mg/day was gradually reduced by 10 mg on every 5 days up to exclusion.

Three months after the last episode she experienced severe, sharp pain in the right side trigeminal nerve innervation (first and second branch) which intensified during speaking, chewing, face washing and touching. The pain lasted for several minutes and reoccurred up to 20 times per day, between attacks she was experiencing a mild dull pain.

Neurological findings: a mild hypoesthesia following the sensory distribution of cranial nerve V, first and second branch. MRI of the brain: aberrant superior cerebellar artery which irritates proximal segments of right trigeminal nerve.

![Figure1. Brain MRI (T2-weighted reconstruction in the coronal plane) showing aberrant superior cerebellar artery which irritates proximal segments of right trigeminal nerve.](image-url)
ebellar artery which irritates proximal segments of right trigeminal nerve (Figure 1). The patient was treated with oxcarbazepine (300 mg bid) for a month after which pain withdrew completely.

Discussion

Cluster headache characterizes recurrence of short, stabbing, unilateral pain in orbital and/or temporal area. It stands for one of the strongest pains one can experience and it can match labour pain, renal colics or fractures [3]. Acute attacks can span from 15–180 minutes and reoccur up to 8 times a day while experiencing autonomic dysfunction symptoms such as lacrimation, nasal congestion, periorbital and conjunctival edema. Opposite to migraine attacks those patients are restless, constantly in motion and not suited by inactivity. The majority of patients (80–90%) has a episodic cluster with acute attacks lasting for 6–12 weeks and remissions up to one year. While durations of acute attacks and remissions can vary significantly among different subjects, they very consistent in the same individuals. First line of treatment during acute attacks consists of the use of oxygen inhalations (100% oxygen 8–10 L/min while standing) and triptans, most commonly subcutaneous sumatriptan injections. For preventive treatment verapamil (240–960 mg/day, tid) and prednisone (60–100 mg per day, for at least 5 days, with gradual discontinuation), when combined are usually beneficial, thus providing usage of lower doses of verapamil compared to therapeutic regime [4]. Trigeminal neuralgia presents as a strong, electrical shock likewise, lancinating pain that is unilateral and usually localized in sensory distribution of one or more trigeminal nerve branches. Acute attack starts abruptly, lasting from several seconds to several minutes and repeats up to couple of hundred times daily. Majority of cases (up to 85%) are episodic and idiopathic in nature with completely normal neurological findings, but with adequate MRI techniques are frequently associated with vascular contact with the trigeminal nerve protuberantial emergence zone. Around 15% of cases are symptomatic with vascular abnormalities in the pontine cistern, with pontocerebellar tumors and multiple sclerosis plaques among commonest causes (dull pain and paraesthesias in cranial nerve V distribution can be reported between attacks) [5]. For pharmacological treatment oxcarbazepine (600–1800 mg/day) and carbamazepine (200–1200 mg/day) represent first line choices. In case of failure, surgical approaches can be considered. Microvascular decompression of a nerve is most efficient option [6, 7].

According to Willbrink et al. [8] the prevalence of trigeminal neuralgia in cluster headache patients is 4.5% (11/244), and it is much higher, opposite to general population (0.1–0.2% or 1–2/1000). As a term cluster-tic was introduced in 1978 [9]. So far there is no open consensus whether it should be recognized as a coexistence of two different types of primary headaches or different phenotype presentation of a unique pathophysiological mechanism. Regardless of underlying pathophysiological mechanism it represents a separate entity consisting of three types of attacks: trigeminal neuralgia attacks, cluster headache attacks and mixed attacks [8]. The International Headache Society in ICHD (International Classification of Headache Disorders, 3rd Edition-beta version) suggest that both of these headaches should receive separate diagnosis and treatments [1].

In our case, attacks of cluster headache and trigeminal neuralgia were typical, occurred in different time periods and had only partially overlapping localization of pain. Separate treatments were introduced, which proved to be very successful. Based on this facts, we believe our patient represents a perfect example of coexistence of two separate entities, cluster headache and trigeminal neuralgia, according to recommendations of International Classification of Headache Disorders, 3rd Edition (beta version).

Competing interests

The authors declare no conflict of interest

References