



The Challenges of Treating Cluster Headache in Peripheral Hospital

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Abstract

Cluster headache is trigeminal autonomic cephalgia with unique pain characteristics with periodicity and autonomic symptoms. Cluster headaches are rare headaches with a higher ratio in males than females. We report a case of cluster headache, a 27-year-old male patient who came in very restless and complained of shortness of breath and severe pain felt on the left side of the face, left ear, left forehead, left eye twitching, blurred vision, and it is challenging to blink, the mouth and left jaw and left nose feel stuffy. On radiological and complete blood count examination did not become the primary support for establishing the diagnosis. Enforcement of the diagnosis and proper treatment comprehensively dramatically affects the patient's prognosis, especially in modifying lifestyle and avoiding triggers of attacks.

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INTRODUCTION

Cluster Headache (CH) is one type of primary headache, which is often called an autonomic trigeminal headache. Other names of cluster headaches are paroxysmal nocturnal cephalalgia, migrainous neuralgia, histamine cephalalgia, red migraine, Horton's headache, and head erythromelalgia.⁷ Cluster headache is a rare case with a prevalence of 1 per 1000 adults in the United States, and other studies have reported 53 per 100,000 adults in 1 year.¹ The incidence of CH is more common in men than women with a ratio of 3 or 4 to 1 and occurs at the age of 20 to 40 years. Pain intensity in CH is usually moderate to severe and is associated with autonomic symptoms.⁹

Based on the International Head Society, the diagnosis of CH include severe unilateral or temporal pain for 15-180 minutes if not treated immediately, headache accompanied by one of the signs of conjunctival injection or lacrimation to ptosis, at the time of the attack, the patient is usually restless. Attacks can be repeated one to eight times a day. Pain is not related to any other disease.¹⁵ Attacks often occur several times at night, especially one and two hours after the patient sleeps.⁷

The prominent symptoms of CH are autonomic symptoms resulting from hyperactivity of the parasympathetic that provide clinical features including ipsilateral lacrimation, red-

ness of eyes, sweaty and nasal congestion. Sympathetic hypoactivity is demonstrated by a combination of ipsilateral ptosis and miosis (droopy eye and smaller pupil on the affected side) and complaints of blurry vision so that the patient comes in a very restless condition.⁹

The management of cluster headaches consists of abortive therapy and prophylactic therapy.¹⁰ This cluster headache has a socio-economic impact related to morbidity; almost 80% of patients reported experiencing limitations in daily activities so that an essential focus in CH management is avoiding triggers and lifestyle modifications.^{1,20}

This case report aims to present the management of rare primary headaches in Indonesia with drug limitations in the peripheral hospital. It was, furthermore, showing the character of autonomic symptoms in CH.

CASE PRESENTATION

At night a 27-year-old man, Javanese, a truck driver, came to the Emergency Room (ER) at Bhayangkara Pusdik Brimob Watukosek Hospital with complaints of severe pain felt on the left side of the face, the pain was felt suddenly for about 1 hour before going to the hospital. The pain is felt like being stabbed from the left ear, left forehead, the left eye feels throbbing with blurred vision and difficulty blinking, left mouth and jaw, and left nose feel stuffy. The

patient came in restless and complained of difficulty breathing and shortness of breath. The pain was felt for 1 hour, so the patient decided to get treatment at the ER. The patient received symptomatic therapy with intravenous ketorolac injection to reduce pain. After the pain subsided somewhat, the patient decided to go home. Thirty minutes later, the patient returned to the ER with the same complaints as before, with a duration of pain felt for 30 minutes. There is a history of previous tooth pain, and no examination or treatment from the dentist has been carried out. There was no history of head injury, history of drug allergy, or disease with similar complaints before, including in the family. Patients often take anti-pain medications in the form of concoctions purchased from herbal shops to relieve complaints of tooth pain. Patients who have a smoking habit in a day can spend 10 to 12 cigarettes.

On physical examination, his blood pressure 160/90 mmHg, heart rate was 86 x/ minute, regular, adequate, respiratory rate was 24 x/minute, symmetrical, regular, temperature axillary 36.6° Celcius and normal oxygen saturation with an intensity of 8/10 in Numeric Pain Rating Scale. On the neurological examination, the patient was vigilant with Glasgow Coma Scale (GCS) E4V5M6. Meningeal sign, motor, and sensory examination were ordinary. On II, III cranial nerve examination was regular. The pupils are isochoric with a positive light reflex,

but the left eyelid looks swollen and feels heavy to blink. We found autonomic symptoms; the patient looks sweaty, has nasal congestion, and is restless. On complete blood count examination, leukocytosis was found ($19.100/\text{mm}^3$), blood sugar was within normal limits, the examination of SARS-CoV2 by rapid antigen test was negative, and chest x-ray results were within normal limits. The results of the dental inspection revealed pulp necrosis, irreversible pulpitis, and chronic apical periodontitis, which required an action plan and dentist treatment.

Initial management in the ER gave 12 liters of oxygen per minute using a non-rebreathing mask showed improvement. Initially, the patient was given an injection of ketorolac (NSAID class) 30 mg intravenously, but the pain only subsided for a moment. In the treatment room, the patient received methylprednisolone 62.5 mg intravenously every 12 hours for three days, ceftriaxone 1 gram intravenously every 12 hours with skin test for three days, and betadine gargle used three times a day. On the second day of treatment, the patient still complained of pain in the teeth with a Numeric Pain Rating Scale of 7/10, so he added an injection of tramadol 100 mg intravenously. The patient was planned for root canal treatment as well as extraction by a conservation dentist. On the third day of treatment, the complaints were much reduced, but sometimes the pain in the teeth was still intermittent. The patient was

allowed to go home by continuing oral therapy with tramadol 100 mg every 12 hours, one teaspoonful of sucralfate syrup three times daily, and multivitamin one tablet orally once a day. One week after the patient came home from the hospital and regularly took the prescribed medication, the headache did not recur, but the teeth' pain sometimes still appears.

DISCUSSION

Cluster headache is one of the primary types of headaches. Primary headaches are caused by spontaneous activation through nociceptive pathways. CH has the characteristics of repeated attacks with moderate to severe pain intensity accompanied by autonomic symptoms, as described in the following International Classification Headaches 3rd (ICHD-III) ^{5,7,9}

Table 1. Diagnostic criteria for cluster headache according to the ICHD III ⁵

A.	At least five attacks fulfilling B through D
B.	Severe or very severe unilateral orbital, supraorbital, and/or temporal pain lasting 15 to 180 minutes if untreated
C.	Headache is accompanied by at least one of the following: Ipsilateral conjunctival injection and/or lacrimation, Ipsilateral nasal congestion and/or rhinorrhea, Ipsilateral eyelid edema, Ipsilateral forehead, and facial sweating, Ipsilateral miosis and/or ptosis, A sense of restlessness or agitation
D.	Attacks have a frequency from one every other day to eight per day
E.	Not attributed to another disorder

The above case fulfills the several diagnostic criteria of the International Classification Headaches in the 3rd edition (ICHD-III). The criteria that fulfill include unilateral pain in the orbital, supraorbital and trigeminal areas. It occurs 1 hour on the first attack, then about 30 minutes on the second attack in a day. Headache followed by nasal congestion, the left eye cannot blink, the vision looks blurry and restless. According to the incidence in male sex and age between 20-40 years and supported by the patient is a heavy smoker.

Cluster headaches consist of episodic and chronic cluster headaches, in which 80% of cases are dominated by episodic cluster headache, the remaining 20% are chronic cases with criteria as shown in table 2:

Cluster headache is a rare headache disorder with a lifetime prevalence of about 0.12%. CH often occurs between the ages of 20 and 40 years and is more common in men with a male-to-female ratio of 3 or 4 to 1.⁹

Table 2. ICHD-III criteria for episodic and chronic cluster headache ⁵

A	Episodic Cluster Headache All fulfilling criteria A through E of Table 1 At least two cluster periods lasting from 7 to 365 days and separated by pain-free remissions of > 1 month
B	Chronic Cluster Headache All fulfilling criteria A through E of Table 1 Attacks recur for > 1 year without remission periods or with remission periods lasting < 1 month.

Cluster headache occurs due to the interaction of three essential structures in the central nervous system, including the trigeminovascular system, autonomic nervous system, and hypothalamus. The hypothalamus has a vital role in pain in CH, where the hypothalamus is activated first, followed by the trigeminovascular and autonomic systems. Furthermore, pain in CH is activated through the trigeminovascular system.⁹

Cluster headache is triggered by risk factors, including smoking lifestyle and alcohol consumption.¹⁸ Almost 70-90% of patients with CH are heavy smokers, both current and past habits. According to the above case, the patient is a heavy smoker. In addition to smoking, drinking alcohol is also a trigger for attacks on CH, where more than 50% of patients who drink alcohol experience attacks on CH. Furthermore, this is probably the basis for the tendency of the male sex to be higher than the female, mainly observed in Asia.¹¹

Based on the case above, signs of infection were also found, as evidenced by routine blood

laboratory results obtained leukocytosis, which is likely the cause of infection from the teeth. In this case, we did not continue with other supporting examinations such as a head CT scan or MRI to rule out other differential diagnoses that lead to secondary headaches because of physical examination. We did not find any abnormalities on neurological examination. In this case, the patient received initial management to give 100% oxygenation of 12 liters per minute. After administering oxygen, the patient felt much-reduced complaints and was not as restless as when he first came. Administration of NSAID anti-pain injections reduces pain only temporarily and still causes a relapse several minutes later. Other therapies include an opioid painkiller, tramadol, a corticosteroid class of methylprednisolone, and a cephalosporin class of antibiotics, which are expected to stop the infection and relieve pain. The therapy given is not under the preparations used to manage acute therapy or abortive therapy for cluster headache because preparations such as sumatriptan, zolmitriptan, or er-

gotamine are not available at our health facilities. However, the patient's progress gradually subsided, and the headache did not recur with oral tramadol opioid painkillers for seven days, but the pain in the teeth still appears sometimes because the patient has not received teeth treatment cause of infection can be eliminated.

Treatment of migraine and cluster headaches includes acute therapy to stop single attacks and prophylaxis therapy from reducing attacks' frequency, duration, and severity. Triptan gives an excellent response to CH attacks, but oral triptan is not recommended because the

effect is longer to stop the attack than giving triptans subcutaneously or intranasally. NSAID is only used as acute therapy in migraines, not in CH.¹⁸

Giving 100% oxygenation with a mask (12-15 lpm flow) is one of the treatment options for CH attacks. A recent randomized controlled trial found that high-flow oxygenated oxygen was significantly more effective than plain air in the acute treatment of migraine and therefore suggested cranial autonomic symptoms or cluster migraine. This response is gotten by inhibiting the activation of nociceptive afferents or autonomic pathways.¹⁸

Tabel 3. Efficacy, Adverse Events, Level of Evidence and Recommendations for the Acute Therapy of Cluster Headache¹⁴

Treatment	Adverse Event	Class	Level
Positive Evidence			
Sumatriptan 6 mg (subcutaneous)	Nonserious: injection site reactions, nausea and vomiting, dizziness, fatigue, paresthesia	I	Level A
Oxygen (100% oxygen 6-12 L/min)	Not reported	I	Level A
Zolmatriptan 5 mg and 10 mg (oral)	Nonserious: paresthesias, heaviness, asthenia, nausea, dizziness, chest pain	I	Level B
Lidocaine 10% (nasal spray)	Nonserious: nasal congestion, unpleasant lidocaine taste	II	Level C
Insufficient Evidence			
Dihydroergotamine 1 mg (nasal spray)	Not reported	III	Level U
Prednisone 30 mg	Not reported	III	Level U

The goal of prophylactic treatment is to reduce the frequency of attacks. However, its continuous use is not recommended because there is no evidence to prevent subsequent attacks. Therefore, the dose of prophylactic therapy can be decreased progressively after two months to 3 months or after the onset of an

attack. The drug is restarted if the attack reappears. If the patient has infrequent attacks, prophylactic therapy may be delayed. In chronic cluster headaches, prophylaxis can be used for a long time, and a low attack rate must be achieved and always consider the potential serious side effects.⁷

Tabel 4 Efficacy, Adverse Events, Level of Evidence and Recommendations for the Prophylactic Therapy of Cluster Headache¹⁴

Treatment	Adverse Event	Class	Level
Positive Evidence			
Verapamil 360 mg daily	Nonserious: constipation, reduced blood pressure, reduced heart rate	II, III	Level C
Lithium 900 mg	Nonserious: polyuria	II	Level C
Melatonin 10 mg every evening	Not reported	II	Level C
Insufficient Evidence			
Capsaicin 0.025% applied twice daily (intranasal)	Not reported	III	Level U
Prednisone 20 mg	Not reported	II	Level U
Negative Evidence			
Sodium valproate 1000-2000 mg daily	Nonserious: nausea and vomiting, somnolence	I	Level B
Sumatriptan 100 mg three times daily	Nonserious: nausea, vomiting, headache, malaise	I	Level B

There are non-invasive and surgical treatments in cluster headache treatment. Neuromodulation such as non-invasive vagus nerve stimulation (nVNS) and radio-surgical treatment is used when oral therapy is not adequate and there are contraindications.¹ Because the pain in CH is one of the most severe pain ever felt

by the patient, the treatment of cluster headache often becomes almost an emergency as patients seek relief. Effective pharmacological therapy options should be available in health facilities to relieve pain during attacks in most CH because cluster headache has a sizeable so-

cio-economic impact, and almost 80% of patients report restricting daily activities. So that the primary modalities in CH include determining diagnosis, prevention of precipitating factors, and adequate therapy can reduce patient morbidity.¹

CONCLUSION

Cluster headaches are a rare case. From this case, we can see that cluster headache is a disease that interferes with daily life and can reduce a person's productivity, especially if they experience repeated attacks and do not get proper treatment immediately. Enforcement of diagnosis to proper treatment comprehensively dramatically influences the patient's prognosis, especially in modifying lifestyle and avoiding triggers of attacks.

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