JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH

How to cite this article :
KUMAR S. DIAGNOSTIC DILEMMA OF PAINFUL RED EYE WITH SEVERE HEADACHE". Journal of Clinical and Diagnostic Research [serial online] 2007 August [cited: 2007 August 1];4:289-292
Available from
http://www.jcdr.net/back_issues.asp?issn=0973-709x&year=2007&month= August &volume=1&issue=4&page=289-292 &id=74

CASE REPORT

Diagnostic Dilemma of Painful Red Eye With Severe Headache

KUMAR S

ABSTRACT

Traditionally, the differential diagnosis of non traumatic painful red eye is limited to microbial keratitis, acute congestive glaucoma & acute iritis. However, if painful red eye is associated with headache, then migraine, sinusitis, orbital infection and inflammation are the usual differential diagnoses. Cluster headache as a cause of painful red eye with severe headache may be missed both by internists as well as by ophthalmologists.

I report two cases initially diagnosed, investigated and treated variously as glaucoma, partial third nerve paresis due to diabetes and meningitis. The correct diagnosis became apparent when all investigations were normal and the patient did not respond to treatment appropriate for the initial, presumed diagnoses. These cases emphasize the importance of a high index of suspicion for cluster headache, and the dramatic response to corticosteroid therapy in this usually overlooked malady.

Key words: severe headache, painful red eye, ptosis

Case Number 1

A 48-year-old female presented with the complaints of recurrent episodes of severe pain in the left eye for last three days. She reported that the pain started suddenly along left supra-orbital margin, within minutes the pain spread in and around eye and became unbearable. Redness of the eye closely followed the onset of pain. The lid became ptotic but the vision remained unaffected.

Injections of Diclofenac sodium were given to relieve the pain. Her past medical and ophthalmic history was unremarkable. She denied any head injury in the past. Investigations revealed that she was an overt diabetic, although her systemic examination was within normal limits ENT examination and X-ray Para-nasal sinus view excluded the sinusitis. A provisional diagnosis of partial third nerve paresis due to diabetes mellitus was made and ophthalmic consultation was advised. Complete blood count, Erythrocyte sedimentation rate, renal function test were within normal limit. VDRL was non reactive and anti nuclear antibodies were not detected. During pain free period the anterior segment was unremarkable except conjunctival congestion. Her intraocular pressure was 32 and 30 mm of Hg. Well defined pink optic disc with CD ratios of 0.7 and 0.6 in the right and left eye respectively excluded the clinical diagnosis of benign intracranial hypertension. Field changes on automated perimetry were consistent with the diagnosis of glaucoma.[Table/Fig 1]. Timolol maleate (0.5%) eye drops, twice a day were prescribed. The intraocular pressure dropped down to 20 mm of Hg but episode of pain recurred and injection Diclofenac was given. Patient was re-evaluated again. Her vision was 6/6 in each eye. There was no proptosis. Although lid edema and mild ptosis was apparent but ocular movements

Corresponding author : Dr Sunil Kumar, FRCS, MS, Mohamad Dossary Hospital, Department of Ophthalmology. PO BOX 335, Al Khobar 31952, Saudi Arabia

were within normal limit. Conjunctiva was congested and chemosed. [Table/Fig 2] Possibility of orbital inflammatory disease or orbital myositis was omitted due to normal ocular movements and absence of proptosis. There was no clinical evidence of scleritis or iritis. Pupil were reactive without an afferent pupillary defect. Gonioscopy revealed grade three open angle in all quadrants. Absence of blood in schlemm's canal excluded the possibility of low-flow carotid-cavernous fistula. Contrast enhanced computerized tomographic (CT) scan of the brain and orbit excluded any structural or space occupying lesion and orbital inflammatory diseases.

Considering the episodic nature of severe periocular pain, conjunctival congestion with mild ptosis, and the normal CT scan findings of the orbit and brain, a presumptive diagnosis of cluster headache was made and treatment was commenced with tablet prednisolone 60 mg per day for three days, and tapered by 10 mg every third day (Campbell protocol). After initiation of this treatment, there was no recurrence of the episodes of pain.

The ptosis as well as conjunctival congestion resolved completely.

Case Number 2:

A 35-year old male patient was brought to the triage with severe headache and pain in the right eye, associated with vomiting, of 30 minutes duration. Although he reported having suffered episodes of headache in the past, but the nature of the current headache was entirely different. Injection Diclofenac sodium was given for a presumed diagnosis of migrainous attack and he was advised to consult the internist in the out patient department. The patient returned in the morning with severe headache, vomiting, painful red right eye and watery rhinorrhea. He was admitted for further evaluation. Systemic examination did not reveal abnormal findings. Results of the complete blood count, erythrocyte sedimentation rate, renal and liver function tests and serum electrolyte levels were found to be within normal limits. The ENT examination was unremarkable. Detailed ocular examination was normal except for the presence of conjunctival congestion. An intracranial space-occupying lesion was excluded since the results of the CT scan of brain were within normal limits. Meanwhile, the patient was treated with intravenous antibiotics,parenteral Metoclopramide, Diclofenac sodium and discharged on third day as headache did not recur.

The patient was re-admitted later the same day as symptom complex recurred. He revealed that the pain had started suddenly along the right superior orbital margin and then spread to the entire head, although it was more intense in the right half. Within minutes, pain was excruciating, the eye became red and he started vomiting. During episode of headache right lid was slightly ptotic although Ocular movements were normal. Conjunctiva was congested and chemosed**.** Mild anisocoria was noted, right pupil was smaller in size. Bilateral fundus examination was normal. Magnetic resonance imaging (MRI) of the orbit and brain to rule out pseudotumor orbit and Tolosa Hunt syndrome were found to be within normal limits.

In view of, excruciating pain around congested right eye along with mild ptosis and miosis, rhinorrhea, unremarkable systemic examination, normal biochemical and radiology profiles, the diagnosis was revised and patient was thought to be suffering from cluster headache. All medicines were stopped and treatment was commenced with oral prednisolone 60 mg per day for three days and tapered by 10 mg every third day. After initiation of the corticosteroids, the headache resolved, conjunctival congestion and ptosis regressed. This patient returned six months later with similar complaints, and responded dramatically to oral prednisolone.

Discussion:

Cluster headache, an uncommon, most painful primary headache, heralds episodes of very severe unilateral orbital, supra-orbital and/or temporal pain, which may extend to the face, neck, ear or hemicranium, with signs and symptoms of autonomic dysfunction. The pain starts without any warning, intensifies rapidly and becomes agonizing in a short time. The designation came from the cluster pattern of attacks, both in terms of the number of episodes per day and the number of days in a bout (cluster pattern)[1]. The frequency of attack may vary from six per 24 hour period to once in a week. If the patient experiences more than three attacks within 24 hours, it may seem that the pain recurs when the effects of analgesics are wearing off. This gives a false impression of

continuous pain, and masks the episodic nature of the illness.

Misdiagnosis of cluster headache is common in clinical practice and can lead to significant morbidity.[2] An interesting non-clinic, population based, internet survey in US observed an average delay of 6.6 years in the diagnosis of cluster headache. The average number of incorrect diagnoses was 3.9 and an average 4.3 physicians were consulted before the correct diagnosis was made, 4% of patients had unnecessary sinus or deviated septum surgery. Many inappropriate medications such as propanolol, amitriptyline, and antibiotics were commonly prescribed while sumatriptan and oxygen were often denied due to a failure to understand the nature of this disorder.[3]

International Headache Society has laid down the criteria for diagnosis of cluster headache. (Table 1: Criteria of international headache society for cluster headache)

At times the clinical boundaries between migraine and cluster headache are ill defined, none meet the strict international society criteria and patient may present with headache sharing the characteristic of both headache type. These occasions are diagnostic challenge.

Change in the pattern or intensity of headache in a patient with history of cluster headache needs neuro-imaging to rule out a structural lesion masquerading as cluster headache. Granulomatous pituitary lesion and intracranial aneurysm may masquerade as cluster headache.[4],[5],[6]

Overlap in symptoms between recurrent orbital myositis and cluster headache may delay the diagnosis of orbital myositis. Orbital myositis should be considered in patients with atypical cluster headache characterized by proptosis, painful eye movements, and pain that does not completely resolve within 3 hours.[7]

Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT syndrome), a newly described headache, characterized by short lasting, unilateral attacks of a neuralgiform pain, accompanied by conjunctival injection and tearing on the symptomatic side, should be considered in the differential diagnosis of cluster headache.

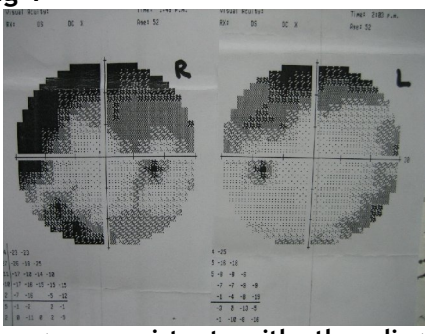
The exact etio-pathophysiological mechanism of cluster headache remains to be elucidated. Recent studies have demonstrated the important role of the ipsilateral ventral hypothalamus in the pathogenesis of cluster headache[8],[9]. The diagnosis of cluster headache is clinical, based on a history of recurrent episodes, the description of the pain, the temporal profile and the accompanying autonomic manifestations. Oxygen inhalation is one of the most effective treatments for cluster headache.[10],[11] Subcutaneous administration of 6 mg of Sumatriptan has been shown to abort the attack of headache in 75 percent of patients[12] but it is contraindicated in patients with hypertension and ischemic heart disease. Sumatriptan is not recommended for patients who experience more than two attacks every 24 hours, in that such use would exceed the recommended limits[13]. In a randomized, placebo controlled, double blind, cross over study, intranasal zolmitriptan 10 mg was found to relieve the headache in 80% of patients with episodic cluster headache[14]. Corticosteroids are the most rapidly acting agents to break the cycle of cluster headache.[13],[15],[16] Kudrow reported significant relief of headache in 77% of 77 patients with episodic cluster headache and partial relief in another 12% of patients given prednisone[17] At the Mayo Clinic, treatment typically is initiated according to the Campbell protocol (i.e., 60 mg daily for 3 days, followed by 10-mg decrements every 3 days over 18 days). Corticosteroids are the drugs of choice in patients with ischemic heart disease or uncontrolled hypertension. If the episodes of cluster are headache are frequent it need prophylactic therapy. Verapamil. lithium, divalproex sodium, and topiramate are effective in the prophylaxis of cluster headache. Recently, Occipital nerve stimulation has been shown to be effective in the treatment of drug-resistant chronic cluster headache.

Conclusion

To conclude, in a patient presenting with excruciating headache of sudden onset, associated with painful red eye and other autonomic dysfunctions, consider the diagnosis of cluster headache. Dramatic relief of the symptom complex by administration of 60 mg of tablet prednisone can be used as a therapeutic test.

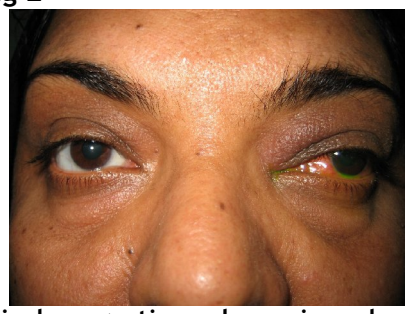
Acknowledgement I wish to acknowledge the precious suggestions and comments of Dr. Philip Thomas in preparation of this manuscript

Table/Fig 1



Field changes consistent with the diagnosis of glaucoma.

Table/Fig 2



Conjunctival congestion, chemosis and mild ptosis in a patient with cluster headache. (photographed with permission from patient)

Table/Fig 3

At least five attacks fulfilling criteria A-C:
A. Severe or very severe unilateral orbital or supra-orbital and or temporal pain lasting 15-180 minutes if untreated.
B. Headache accompanied by at least one of the following: 1. Ipsilateral conjunctival injection and/or lacrimation 2. Ipsilateral nasal congestion and/ or rhinorrhea 3. Ipsilateral eyelid edema 4. Ipsilateral forehead and facial sweating 5. Ipsilateral ptosis and miosis 6. A sense of restlessness and agitation
C. Attacks occur in a frequency of one every other day to eight per day
D. Pain not attributable to any other disorder.

International Headache Society criteria for diagnosis of cluster headache

References:

[1] Kunkle E, Pfeiffer J, Wilhoit W. Recurrent brief headaches in "cluster" pattern. Trans Am Neurol Assoc 1952;77:240-243
[2] Geweke LO. Misdiagnosis of cluster headache. Curr Pain Headache Rep 2002;6:76-82.
[3] Klapper JA, Klapper A, Voss T. The misdiagnosis of cluster headache: a nonclinic, population-based, internet survey. Headache 2000;40:730-5.
[4] Favier I, Haan J, van Duinen SG, Ferrari MD. Typical cluster headache caused by granulomatous pituitary involvement. Cephalalgia 2007;27:173-6.
[5] Gentile S, Fontanella M, Giudice RL, Rainero I, Rubino E, Pinessi L. Resolution of cluster headache after closure of an anterior communicating artery aneurysm: the role of pericarotid sympathetic fibres. Clin Neurol Neurosurg 2006;108:195-8.
[6] McBeath JG, Nanda A. Case reports: sudden worsening of cluster headache: a signal of aneurismal thrombosis and enlargement. Headache 2000;40:686-8.
[7] Lee MS, Lessell S. Orbital myositis posing as cluster headache. Arch Neurol 2002;59:635-6.
[8]Goadsby PJ. Cluster headache: New perspectives. Cephalalgia 1999;19:39-41
[9] May A, Leone M. Update on cluster headache. Curr Opin Neurol 2003;16:333-340
[10] Kudrow L. Response of cluster headache attacks to oxygen inhalation. Headache 1981;21:1-4
[11] Fogan L. Treatment of cluster headache: a double-blind comparison of oxygen v air inhalation. Arch Neurol 1985;42:362-363
[12] Treatment of acute cluster headache with sumatriptan. The Sumatriptan cluster headache Study Group. N Engl J Med 1991;325:353-354
[13] Silberstein SD. Pharmacologic management of cluster headache. CNS Drugs 1994;2:199-207
[14] Cittadini E, May A, Straube A, Evers S, Bussone G, Goadsby PJ. Effectiveness of intranasal zolmitriptan in acute cluster headache: a randomized, placebo-controlled, double-blind crossover study. Arch Neurol 2006;63:1537-42.
[15] Jammes JL. The treatment of cluster headaches with prednisone. Dis Nerv Syst 1975;36:375-376
[16] Couch JR, Ziegler DK. Prednisone therapy for cluster headache. Headache 1978;18:219-221
[17] Kudrow L. Cluster Headache: Mechanisms and Management. Oxford University Press New York:1980