Orbital Myositis Posing as Cluster Headache

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Objective: To describe the case of a patient with recurrent orbital myositis who was thought to have cluster headaches for 6 years.

Design and Setting: Case report in an outpatient neuro-ophthalmology clinic.

Patient: A 24-year-old man developed unilateral supraorbital pain, lacrimation, conjunctival hyperemia, nasal congestion, proptosis, and painful eye movements. The pain intensity varied over the course of each day and disappeared after 1 month. He had multiple attacks responsive to prednisone that were separated by months over the ensuing 6 years. Neuroimaging revealed an enlarged extraocular muscle.

Conclusions: Overlap in symptoms between recurrent orbital myositis and cluster headache delayed the diagnosis in this patient. Orbital myositis should be considered in patients with atypical cluster headache characterized by proptosis, painful eye movements, and pain that does not completely resolve after 3 hours.

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THE INTERNATIONAL Headache Society has established criteria for the diagnosis of cluster headache (Figure 1).1 It predominantly affects men between 20 and 40 years of age and centers in and around 1 eye. Episodes begin acutely, last 1 to 2 hours, and occur several times a day. They occur daily for an average of 2 weeks to 3 months (International Headache Society criteria are 1 week to 1 year). Associated findings include ipsilateral conjunctival hyperemia, lacrimation, ptosis, miosis, nasal stuffiness, and facial sweating.2

Recurrences almost always occur on the same side, but 10% to 15% may alternate. Remissions should last at least 2 weeks, but they often will last months to years. About 10% of patients suffer chronic cluster headaches, with “clusters” occurring for more than 1 year without remission or with remissions lasting less than 2 weeks.1 Orbital myositis is a subgroup of idiopathic orbital inflammatory syndrome. It is the second most common cause of extraocular muscle enlargement, after thyroid-associated ophthalmopathy. Women are affected twice as often as men, and the mean age of onset is approximately 30 years. Mombaerts and Koornneef3 followed up 16 patients with myositis for a mean of 9.7 years and found 56% with recurrence. Clinical findings include painful eye movements, lid edema, conjunctival hyperemia and chemosis, poor extraocular motility, and proptosis.

We present the case of a patient with recurrent orbital myositis, which was mistakenly diagnosed and treated as cluster headache for 6 years.

REPORT OF A CASE

In 1994, a 24-year-old man developed paroxysmal headache in the right supraorbital region associated with swelling of his right eyelid and eye. The headache was severe, pounding, and not preceded by an aura. The patient also noted blurring of the vision in his right eye and painful eye movements. The headache fluctuated in intensity over the course of the day and lasted approximately one month. Recurrences were separated by 2 months and precipitated by alcohol use. He denied having sinus disease. Apart from 2 uncles with migraine, his family history was noncontributory.

Results of ophthalmologic evaluation for proptosis between episodes were reportedly normal. A computed tomographic scan of the brain and orbits dur-
ing an attack demonstrated enlargement of the right medial rectus muscle. Medical history was unremarkable. Treatment with over-the-counter nonsteroidal anti-inflammatory medicine and acetaminophen/butalbital/caffeine gave only minor relief.

In 1996, neurologic consultation revealed tenderness to palpation of the right eye with fullness of the right upper lid, conjunctival hyperemia, lacrimation, and nasal stuffiness. The results of a pupil examination were normal, and there was no optic nerve edema. The neurologic examination was otherwise unremarkable. Episodic cluster headache was diagnosed, and the patient was treated with prednisone. The results of thyroid function tests and erythrocyte sedimentation rate were normal.

The headaches recurred in the spring and fall of each year for the next 5 years, each time on the right side with a good response to prednisone. In August 2000, the patient developed left retro-orbital headache with blurred vision and lacrimation of the left eye associated with proptosis and ptosis of the left eye with conjunctival hyperemia, and nasal stuffiness sometimes precipitated by the ingestion of alcohol. As in some cases of cluster headache, the disorder was responsive to corticosteroids. The correct diagnosis is established when imaging of the orbit demonstrates enlargement of an extraocular muscle. It is important that orbital pseudotumor be diagnosed expeditiously because prompt treatment with oral corticosteroids is apt to provide quick relief and prevent reduced ocular motility. Furthermore, an inflamed orbit may be a manifestation of orbital cellulitis or a systemic disorder such as Wegener granulomatosis. In light of this, it behooves neurologists to consider obtaining either computed tomographic scans or magnetic resonance images of the orbits in cases of cluster headache in which eyelid edema, ptosis, and conjunctival hyperemia are prominent features or in which the duration of pain is prolonged, as with our patient.

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REFERENCES


Cluster headache, a disorder of unknown cause and pathogenesis, can be mimicked by other disorders. Instances in which vertebral artery aneurysm, cavernous sinus pseudoaneurysm, and arteriovenous malformations were confused with cluster headache have been reported. It is easy to see why, as in our case, inflammatory orbital pseudotumor could be confused with cluster headache. The patient was a young man with recurrent, unilateral, high-intensity pain accompanied by eyelid edema, lacrimation, ocular hyperemia, and nasal stuffiness sometimes precipitated by the ingestion of alcohol. As in some cases of cluster headache, the disorder was responsive to corticosteroids. The correct diagnosis is established when imaging of the orbit demonstrates enlargement of an extraocular muscle. It is important that orbital pseudotumor be diagnosed expeditiously because prompt treatment with oral corticosteroids is apt to provide quick relief and prevent reduced ocular motility. Furthermore, an inflamed orbit may be a manifestation of orbital cellulitis or a systemic disorder such as Wegener granulomatosis. In light of this, it behooves neurologists to consider obtaining either computed tomographic scans or magnetic resonance images of the orbits in cases of cluster headache in which eyelid edema, ptosis, and conjunctival hyperemia are prominent features or in which the duration of pain is prolonged, as with our patient.