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A Painful Ophthalmoplegia- Tolosa Hunt Syndrome: A Case Report

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Abstract

Tolosa-Hunt Syndrome is an uncommon condition characterized by painful ophthalmoplegia affecting the third, fourth, and/or sixth cranial nerves, which is caused by an unknown etiology of non-specific inflammation in the cavernous sinus or superior orbital fissure. a 60-year-old diabetic male patient who recovered from covid-19, 15 days before, came with complain of sharp and intense unilateral headache, ptosis, ophthalmoplegia and loss of vision in right eye in the past 10 days diagnosed with Tolosa-Hunt Syndrome was presented. He was suffering from a right-sided headache, periorbital pain, and double vision. Right-sided ptosis, right-sided trochlear and abducens nerve palsy, and partial right-sided oculomotor nerve palsy with hypoesthesia in the ocular division of the trigeminal nerve were discovered during the examination. The optic nerve and lateral rectus muscle revealed abnormal signal intensity variations on magnetic resonance imaging showed the minimal proptosis noted on the right side with mild edema in the right periorbital soft tissues and small enhancing soft tissue in the right cavernous sinus region which suggested the possibility of Tolosa Hunt Syndrome of the head and orbit. Pain and ptosis were greatly reduced after 72 hours of steroid treatment. Tolosa-Hunt Syndrome is an exclusionary diagnosis with a clinical presentation, normal tests, and magnetic resonance imaging. Recurrent unilateral orbital pain, ipsilateral oculomotor paralysis, and a quick reaction to steroids characterize Tolosa–Hunt syndrome, a severe ophthalmoplegia Treated with intravenous methyl prednisolone (40 mg) once a day with other supportive care therapy. Yet, there was no reversal of vision loss and ptosis. However, the headache subsided and the patient was discharged after 7 days. In Vadodara, Gujarat, India this is most likely the first occurrence and reported case of Tolosa–Hunt syndrome.

Keywords: Painful ophthalmoplegia, intense unilateral headache, Tolosa-Hunt Syndrome

Introduction:

Tolosa-Hunt syndrome (THS), also known as painful ophthalmoplegia, recurrent ophthalmoplegia, or ophthalmoplegia syndrome, is described as severe and unilateral periorbital headaches associated with painful and restricted eye movements. THS is one of the rare disorders recognized by the National Organization for Rare Disorders.[1] It is also included as one of the painful cranial neuropathies by the International Headache Society (HIS) in its headache classification. Tolosa Hunt syndrome is usually idiopathic and is thought to be from non-specific inflammation in the region of the cavernous sinus and/or superior orbital fissure. However, traumatic injury, tumors, or an aneurysm could be the potential triggers. The annual estimated incidence of THS is about one case per million per year. It is caused by non-specific inflammation of unknown etiology. It is usually diagnosed via exclusion, and as such a vast amount of laboratory tests are required to rule out other causes of the patient's symptoms. These tests include a complete blood count, thyroid function tests and serum protein electrophoresis. Studies of cerebrospinal fluid may also be beneficial in distinguishing between THS and conditions with similar signs and symptoms. Treatment includes immunosuppressive such as corticosteroids (methylprednisolone) and steroid-sparing agents like methotrexate or azathioprine.

Patients with headaches are frequently seen by primary care physicians, nurse practitioners, and internists. When they encounter a patient with a severe headache and ophthalmoplegia, they must report them to a neurologist. Patients with THS are treated with steroids and immunosuppressive drugs, rather than the usual headache medication. Recurrences are prevalent despite treatment, and overall quality of life is low. When patients are given steroids, the nurse practitioner and primary care provider should keep an eye on them for any negative side effects. The optimal patient outcome will come from an interdisciplinary approach to this disease's treatment.

Case Description:

A 60-year-old diabetic male patient who recovered from covid-19 15 days before, came with complain of sharp and intense unilateral headache, ptosis, ophthalmoplegia and loss of vision in right eye in the past 10 days. He was admitted to another hospital where he was diagnosed with THS by MRI Brain and other laboratory tests. He was advised to start intravenous corticosteroid methyl prednisolone once a day for seven days; however, he took discharge against medical advice after 1st dose. After 5 days, the symptoms intensified and so he came to our hospital. Later, again his MRI study of brain with orbit was done which showed the minimal proptosis noted on the right side with mild edema in the right periorbital soft tissues and small enhancing soft tissue in the right cavernous sinus region which suggested the possibility of THS. The pharmacotherapy involved intravenous methyl prednisolone (40 mg) once a day with other supportive care therapy. Yet, there was no reversal of vision loss and ptosis. However, the headache subsided and the patient was discharged after 7 days. The discharge medication included Tab. Methylprednisolone (16 mg) [2-0-1] for 5 days then taper to [1-0-1] for 5 days, Cap. Pantoprazole (40 mg) [1-0-0] and Tab. Paracetamol (650 mg) [SOS].

Discussion:

THS is also known as painful ophthalmoplegia, recurrent ophthalmoplegia, and ophthalmoplegia syndrome THS is a rare disorder characterized by severe periorbital headaches, double vision, paralysis (palsy) of certain cranial nerves, and chronic fatigue. Affected individuals may also exhibit protrusion of the eye (proptosis), drooping of the upper eyelid (ptosis) double vision (diplopia), large pupil, and facial numbness with diminished vision. In most cases, symptoms associated with THS affect only one side (unilateral). Symptoms will usually subside without intervention (spontaneous remission) and may recur without a distinct pattern (randomly) along with decreased and painful eye movements (ophthalmoplegia). Symptoms usually affect only one eye (unilateral). In most cases, affected individuals experience intense sharp pain and decreased eye movements. Symptoms often will subside without intervention (spontaneous remission) and may recur without a distinct pattern (randomly). The exact cause of THS is not known, but the disorder is thought to be associated with inflammation of specific areas behind the eye (cavernous sinus and superior orbital fissure).[1] in some cases of severe ophthalmoplegia, the eye itself is unable to move or look in various directions (frozen globe). This disease is caused by an unspecific inflammation in the region of the cavernous sinus and the sphenoid cleft. Its evolution is considerably shortened by corticosteroid therapy correctly within 72 hours.[2]. It is a diagnosis of exclusion retained after eliminating other etiologies of painful ophthalmoplegia.[3, 4] THS can affect people of any age, without predominance of sex. In general, it is unilateral, but cases of bilaterality have been reported.[5] the diagnosis of this syndrome was retained in the face of the negativity of all the investigations carried out as well as the response to treatment the diagnosis of THS has been codified by the International Headache Society. The following criteria must be met for a definitive diagnosis of this disorder: Eye pain on one side of the head that persists for at least eight weeks if untreated; associated irritation or damage to the third, fourth, or sixth cranial nerves; relief of pain within 48 hours upon the administration of steroids; and specialized testing that rules out other conditions such as neoplasm, infection or aneurysm. The diagnosis of THS is suspected based upon the presence of characteristic physical features (e.g., pain, headache, ophthalmoplegia). The diagnosis may be confirmed by a thorough clinical evaluation, detailed patient history, and a variety of specialized radiologic tests including computed tomography (CT) scan, and magnetic resonance imaging (MRI). These examinations may reveal characteristic enlargement or inflammation of the areas behind the eye (cavernous sinus and superior orbital fissure).[1] CBC count, erythrocyte sedimentation rate (ESR), electrolytes with glucose, thyroid function tests, fluorescent treponemal antibody (FTA), antinuclear antibody (ANA), lupus erythematosus (LE) preparation, antineutrophil cytoplasmic antibody (ANCA), serum protein electrophoresis, Lyme titre, angiotensin-converting enzyme (ACE) level, and HIV titre are helpful in eliminating other processes. This level of evaluation is required to exclude other conditions, which can have significant morbidity associated. Cell count and differential, protein, glucose, fungal and/or bacterial cultures, Gram stain, cytology, and opening pressure of CSF are helpful in eliminating conditions mimicking THS; a mild (lymphocytic) pleocytosis within the spinal fluid may occur in patients with THS. Anti-GQ1b antibodies may be helpful in distinguishing early, painless THS from Miller Fisher syndrome. MRI of the brain and orbit with and without contrast, magnetic resonance (MR) angiography or digital subtraction angiography (DSA), and CT

scan of the brain and orbit with and without contrast may all be useful (see the images below). Inflammatory changes in the cavernous sinus, superior orbital fissure, and/or orbital apex are typically observed on high resolution contrast enhanced imaging. In the authors' experience, thin-slice high–magnetic field MRI of the cavernous sinus region, including coronal sections with and without contrast and fat-suppressed cuts of the orbital regions, is the modality of choice. These changes are not specific for THS and may also be present in neoplastic conditions of the cavernous sinus. Enlargement of the optic nerve or external ocular muscles has been described, emphasizing the continuum with idiopathic orbital inflammatory disorders.[6]

Etiology and Prognosis:

While the exact cause of THS is unknown, one theory is an abnormal autoimmune response linked with an inflammation in a specific area behind the eye (cavernous sinus and superior orbital fissure). In some cases, inflammation may be due to a clumping of a certain type of cell (granulomatous inflammation). Autoimmune disorders are caused when the body's natural defenses against "foreign" or invading organisms (e.g., antibodies) begin to attack healthy tissue for unknown reasons. Other possible causes may include generalized inflammation and constricted or inflamed cranial blood vessels. The substantial clinical improvement seen with glucocorticoids is a hallmark of THS. Symptom alleviation, particularly pain relief, usually occurs within 24 to 72 hours of commencing steroids, with the majority of patients reporting improvement within a week. Cranial nerve palsies improve with time, and recovery might take anywhere from two to eight weeks.[7] after steroid treatment, persistent impairments are uncommon. Relapses happen in roughly 40% to 50% of patients, and they can be ipsilateral, contralateral, or bilateral.[8] Younger people are more likely to relapse than older patients. Because THS is an exclusionary diagnosis, every relapse should preferably be examined with a thorough workup.[9] Steroids aren't known to assist people avoid relapses.[10]

Affected Population: THS is a rare neuro-immunological condition that affects both men and women equally. Although the typical age of onset is 41 years, instances have been observed in people as young as 30 years old. THS has been diagnosed in children under the age of ten in rare circumstances.

Differential Diagnosis: The following conditions have symptoms that are comparable to THS. For differential diagnosis, comparisons may be useful like the inflammation of the tissues within the cavity that contains the eyeball is known as orbital cellulitis. Extreme pain, impaired eye movement, swelling, fever, and a general sensation of discomfort are all symptoms. Impaired vision, vascular abnormalities, and inflammation spreading to the entire orbit, brain, or membranes around the brain are all possible problems. Cavernous sinus thrombosis is an ophthalmologic condition characterized by infection and clotting in veins behind the eyes. It might be a side effect of orbital cellulitis or a skin infection on the face. Both eyes are swollen and protruding, there is a fever, a headache, impaired eye movements, droopy eyelids, facial numbness, and a sickly appearance.

Standard treatment as per guideline: In most cases, the pain associated with THS subsides with short-term use of steroid drugs. Pain is usually reduced in untreated cases within fifteen to twenty days. With steroid treatment, pain typically briskly subsides within twenty-four to seventy-two hours and this brisk steroid response aids in the diagnosis. Affected individuals may be vulnerable to recurrent future attacks.[11]

Treatment plan for this patient: Glucocorticoids have been the mainstay of the treatment ever since the syndrome was first described. But there is no specific data to give recommendations about dose, duration, or route of administration. Spontaneous remission of symptoms is known to occur. Although orbital pain drastically improves with steroid treatment, there is no evidence to suggest cranial nerve palsies improve faster with it. As with any glucocorticoid regimen, treatment for THS involves initial high-dose therapy for few days followed by a gradual taper over weeks to months. A very small percentage of patients will require immune suppression with other agents, either to avoid side effects of long-term steroid therapy or for long-term suppression of the disease process itself. Azathioprine, methotrexate, mycophenolate mofetil, cyclosporine, and infliximab have been used as second line therapy.

Conclusion:

Unlike the typical headache, patients with the THS are managed with steroids and immunosuppressive agents. Despite treatment, recurrences are common and the overall quality of life is poor. When patients are managed with steroids, the nurse

practitioner and primary care giver should monitor the patient for adverse effects. THS is a rare condition with a well-defined clinical picture but no recognized cause. It's still an exclusionary diagnosis. The lack of positive results from studies and a remarkable reaction to corticosteroids allowed us to keep the diagnosis in our case. An inter-professional approach to the care of this disease will provide the best patient outcome.

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