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# Tolosa hunt syndrome: case report of a 50-year-old man with unilateral headache ophtalmoplegia

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**ABSTRACT**
Tolosa Hunt Syndrome (THS) is a rare clinical syndrome described as unilateral orbital or periorbital pain involving parese of one or more cranial nerves oculomotor (III), trochlear (IV), and or abducens (VI) caused by nonspecific granulomatous inflammation on the cavernous sinus, superior orbital fissure or orbital. MRI images can show thickening at cavernous sinus or orbital fissure. Giving corticosteroids for 48 hours resulted improvement in clinical symptoms. This article is a case report of a 50 years old man with Tolosa Hunt Syndrome. A 50-year-old man has come with complaints of recurrent headaches for 6 months in the left eye area, pain when moving his eyes, accompanied by burning sensation like electrocution, with difficulty moving the left eyeball to the left side and diplopia. Laboratory tests was normal, on MRI examination there is thickening of the musculus rectus lateralis. The patient was given injection methylprednisolone 125 mg/8 hours and gave good responses. Diagnosis of tolosa hunt syndrome (THS) in this case enforced based on the presence of unilateral headache in the periorbital area accompanied by weakness of the optic nerve (cranial nerves III and VI left), burning and electrifying feeling in the unilateral facial area indicating involvement of the nerve V1. Chronic inflammation of the cavernous sinus wall septa in dense connective tissue can exert pressure that can penetrate cranial nerves III, IV, VI and V branch 1. By giving of systemic corticosteroids for 48 hours resulted in improvement in clinical symptoms.



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## INTRODUCTION

Tolosa Hunt Syndrome (STH) is a syndrome characterized by recurrent unilateral headaches accompanied by painful ophthalmoplegia and eye movement disorders. This disease is recognized by the National Organization for Rare Disorders (NORD) as a rare disease. Tolosa Hunt syndrome was first described by Dr. Eduardo Tolosa in 1954 a Spanish neurosurgeon. A similar case was reported by Hunt et. al in 1961, and in 1966 Smith and Taxdal used the term Tolosa Hunt Syndrome for this disease for the first time (Patel et al., 2015).

The annual incidence is estimated at 1:1000,000, usually occurring in people aged 40 years and over, women and men have the same chance of getting this disease (Patel et al.,

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2015). According to the International Headache Society (IHS), Tolosa Hunt Syndrome is defined as unilateral orbital or periorbital pain associated with paresis of one or more of the oculomotor (III), trochlear (IV), and abducens (VI) cranial nerves caused by granulomatous inflammation of the sinuses. cavernous, superior orbital or orbital fissure (HIS, 2016)

The clinical manifestations of Tolosa Hunt Syndrome are typical symptoms in the form of pain with a severe sharp or stabbing sensation, which can come and go and occurs quite frequently. Pain usually occurs in the periorbital area but can also be retro orbital and can spread to the frontal and temporal areas and is felt unilaterally. Pain in the eyes can be accompanied by ophthalmoplegia which occurs simultaneously or gradually from the initial onset of pain (Somlai et al., 2016). Some found nausea and vomiting possibly due to the severe pain experienced and also chronic fatigue (Amrutkar, 2020). Other complaints such as diplopia or double vision, exophthalmos or mild eye protrusion and eye movement disorders such as difficulty moving in certain directions can also be found (Somlai et al., 2016)

Tolosa Hunt syndrome is caused by nonspecific granulomatous inflammation of unknown etiology. Chronic inflammation of the septa of the cavernous sinus wall in dense connective tissue can exert pressure that can penetrate cranial nerves III, IV, VI and V branches 1 (Amrutkar, 2020). Because the cavernous sinus is inflamed, the three nerves that move the eyeball which run together in the cavernous sinus pathway can experience paresis (Somlai et al., 2016).

Paraesthesia of cranial nerve V branch 1 indicates an inflammatory processintracavernous. Involvement of cranial nerve V branch 1 and maxillary branch (V branch 2) indicates that inflammation can progress anteriorly until it reaches the superior orbital fissure. If inflammation extends superomedially to reach the apex of the orbit, it results in disruption of cranial nerve II. 5 In orbital localization, compression of cranial nerve II occurs due to intracavernous inflammation causing loss of vision (Somlai et al., 2016)

MRI or CT-Scan examinations can be used to detect inflammation in the cavernous sinus or orbital fissure (Amrutkar, 2020). MRI of the head, especially the coronal section, is the main imaging in establishing a diagnosis of STH.5 Blood and CSF examinations are expected to be normal in cases of STH, although in some cases there is an increase in ESR and leukocyte count (Somlai, 2016). If any abnormalities are found, then other diagnoses must be considered. (Amrutkar, 2020) Lumbar puncture examination is also an examination to rule out the possibility of bacterial or viral infections. In STH the results were normal, although there was a slight increase in protein and cells that responded to corticosteroid administration. (Sumanungkalit et al., 2019)

The diagnosis of Tolosa Hunt Syndrome is based on diagnosis of exclusion. This disease is considered after excluding other causes by clinical, laboratory and imaging examinations, and the patient responds well to corticosteroid administration. The diagnostic criteria for Tolosa Hunt Syndrome based on the International Headache Society in 2018 are as follows: 1)Unilateral orbital or periorbital headache according to criterion C, 2)These two things:(a)Granulomatous inflammation of the cavernous sinus,fissuresuperior orbital based on MRI examination or biopsy, (b) Paresis of one or more of cranial nerves III,

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IV and/or VI.1)Proof of causation is demonstrated by both of the following:(a)Ipsilateral headache due to granulomatous inflammation. (b)Headache followed by the development of paresis of nerves III, IV, and/or VI within a period of ≤ 2 weeks. 2)Not better accounted for by other ICHD-3 diagnoses.

There is a time span between the onset of pain in the eye area and its onsetophthalmoplegiaalso considered as STH (Samatra, 2016) Administration of systemic corticosteroids to STH for 48 hours results in improvement of clinical symptoms so that it can help rule out the differential diagnosis.

The most common complications are side effects of long-term use of high-dose corticosteroids. Tolosa Hunt syndrome is often a harmless condition but some cases involve cranial nerve II and in severe cases can cause blindness (Lasam et al., 2016) Some cases that respond well to corticosteroid administration have a good prognosis. Accurate diagnosis as early as possible can reduce the duration and severity of STH symptoms such as vision loss (Filho, 2016)

## METHODS

This type of research is case study research oris a case report of a 50 year old man with Tolosa Hunt Syndrome. A 50 year old man presented with complaints of recurring headaches for 6 months in the left eye area, pain when moving the eyeball, accompanied by a burning sensation like an electric shock, difficulty moving the left eyeball to the left side (ophthalmoplegia) and double vision. Laboratory examination showed normal results, on MRI examination there was thickening of the lateral rectus muscle.

## RESULTS AND DISCUSSION

### Results

A 50 year old man came to the emergency room at the Praya Regional General Hospital in a conscious state complaining of headachesfeltsince 6 months ago it has been coming and goingburdensomesince 2 days ago. Headache is felt in the left eye area, pain when moving the eyeball, accompanied by a burning sensation like an electric shock on the face. Headache appears suddenly, does not get worseactivityand it doesn't get better with rest. The patient also complained that the left eyelid could not open and close properly, accompanied by difficulty moving the eyeball to the left side and felt a little protruding since 2 days ago, apart from that the patient complained of double vision. The patient also complained of vomiting. During these 6 months of complaints of intermittent pain, the patient only took the anti-pain medication paracetamol or ibuprofen which he bought himself at the pharmacy. The patient denies having a fever, does not have a cough, cold or diarrhea and has never experienced itinjuryprevious head, complaints of fainting were also denied. When this pain appears, the patient does not complain of tears coming out of his eyes, nor does he complain of excessive sweating or dizziness. The patient has no history of trauma, history of diabetes and no history of high blood pressure so far.

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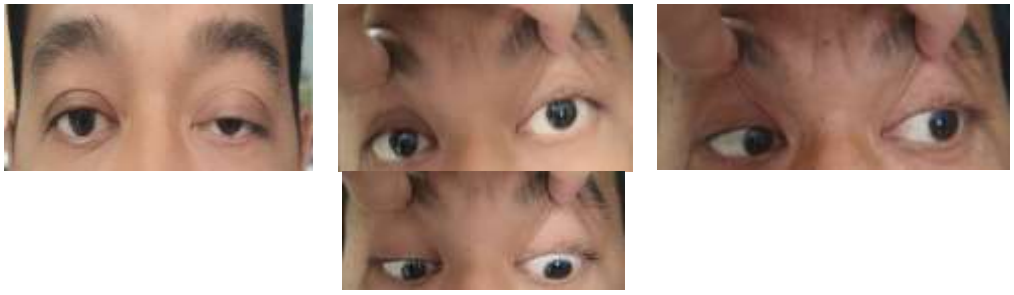

**Figure 1.** Ptosis, protopsis and paresis of left III and VI nerves are visible

Patientdiagnosedwith susp. Tolosa Hunt syndrome andplannedHospitalization to carry out routine blood tests, kidney function blood sugar, EKG and head CT scan without contrast. While in the emergency room, the patient received injection therapyomeprazole 40 mg/12 hours, inj.Ondansetron4mg/8 hours, inj.Methylprednisolone 125 mg/8 hoursas well as oral analgesics 1 tab/8 hours and flunarizine 1 tab/12 hours.ondansetroninjection 4mg/8 hours, carbamazepine 200mg/12 hours. Laboratory examination showed a normal impression (6.2x/µL). When checking blood sugar, it was found to be 92 mg/dL, with urea and creatinine within normal limits (20.5 mg/dL and 0.66 mg/dL). ECG examination interpretation is normal sinus rhythm. CT scan of the head without contrast was within normal limits.$\mathbf{10^3}$

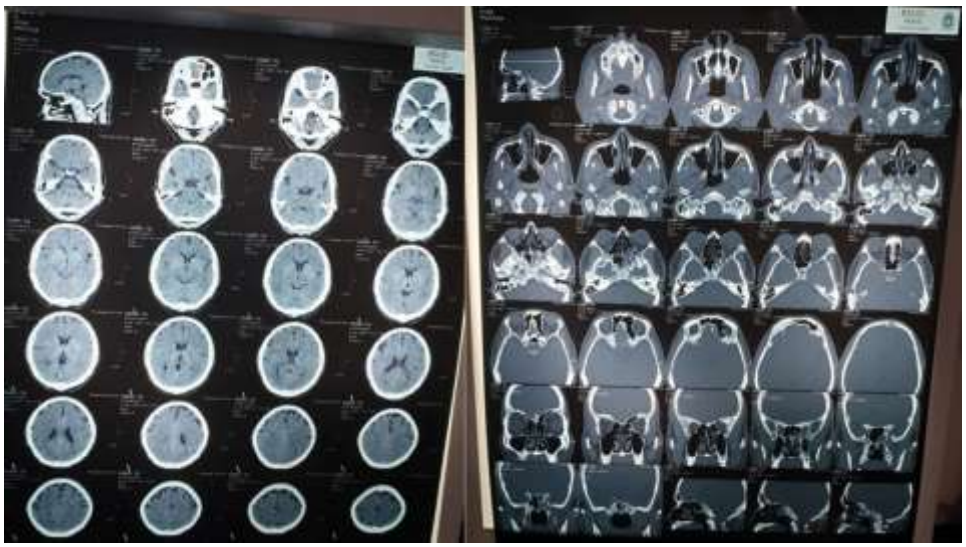

**Figure 2.** Ct scan of the head without contrast shows normal results

After 2 days of treatment the patient showed a positive response to treatment which was marked by reduced pain, tingling and electric shock sensationsin the facealso improved, double vision gradually returned to normal. Patientallowedgo home andplannedhead MRI examination at the referral hospitalafterthe control came back 5 days later. The therapy that the patient took home was methylprednisolone 3x16mg and mecobalamin 2x500mg.

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At the next visit to the neurology clinic, the patient brought the MRI results from the referral hospital. The patient's complaints also began to improve, the pain was much reduced, the ptosis improved, double vision reduced, and eye movement also improved. MRI results were obtained with the impression of thickening of the lateral rectus muscle.

**Figure 3.** Clinical improvement of ptosis and movement of the left eyeball.

**Figure 4.** Head MRI results showed an impression of thickening of the lateral rectus muscle

## Discussion

In this patient the diagnosis was STHenforcedbased on the presence of unilateral headache in the periorbital area accompanied by weakness in the nerves that move the eyeball (cranial nerves III and VI left). Patients also complain of burning and electric shocksin areaunilateral facial involvement, indicating involvement of cranial nerve V1.Tolosa Hunt syndrome is caused by granulomatous inflammationnonspecificof unknown etiology.4 Chronic inflammation of the septa of the cavernous sinus wall in dense connective tissue can exert pressure that penetrates the branches of cranial nerves III, IV, VI and V. 1.4 Because the cavernous sinus is inflamed, the three nerves that move the eyeball

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run together. in the cavernous sinus pathway, paresis can occur. 3 Paraesthesia of cranial nerve V branch 1 indicates an intracavernous inflammatory process. (Luminatobing, 2018)

Cranial nerve III paralysis can cause ptosis, namely drooping eyelids, as in the patient in the case. This is due to paralysis of the levator palpebral muscle. Mild paralysis if one eye slit is smaller and the forehead is wrinkled to compensate. While total paralysis is found the eyelids cannot be opened (Luminatobing, 2018).

Diplopia is caused by paralysis of one of the nerves that move the eyeball, namely cranial nerve III, which causes a "down and out" position in the affected eye, as experienced by the patient in this case (Olse et la., 2017).

Examinations related to cranial nerve II such as visual acuity, visual field, and funduscopic examination of the papillae, retina and macula are also needed if the patient complains of decreased vision. Case report Simanungkalit et. al stated that STH involving cranial nerve II was found in one eye with 0 visual acuity, the appearance of hyperemic papillae with indistinct boundaries and an artery:vein ratio of 1:3 consistent with papilledema (Sumanungkalit et al., 2019).

In this case the patient experienced unilateral pain that came and went for 6 months, and in the last 2 days it got worse accompanied by movement disorders of the left eyeball.There is a time span between the onset of pain in the eye area and its onsetophthalmoplegiaalso considered as STH (Samatra et al, 2016).

The nausea and vomiting experienced by the patient in this case was due to the severe pain experienced and chronic fatigue was also found.In this patient laboratory examination was found to be normal.Although in cases of STH it is expected to be normal, in some cases of STH there is an increase in ESR and leukocyte count without a clear focus of infection. 3 Lumbar puncture examination is also one of the examinations to rule out the possibility of bacterial or viral infection. Normal results support STH, although in some cases there is a slight increase in proteins and cells that respond to corticosteroid administration (Kline, 2001).

The results of the patient's MRI imaging in the case showed an impression of thickening of the left lateral rectus. The imaging features of STH are not specific. Both MRI and CT scans can be used to detect inflammation of the cavernous sinus or orbital fissure and thickening of the cavernous sinus (Sumanungkalit et al., 2019).

The differential diagnosis for eye pain and ophthalmoplegia is infection, orbital cellulitis, sinus thrombosiscavernous, neoplasms, vascular disorders and autoimmune diseases. Cavernous sinus thrombosis can be septic or non-septic. Patients often come with complaints of acute fever, proptosis, ptosis,chemosis, periorbital edema, headache, optic disc edema,escophthalmusand paralysis of cranial nerves III, IV, and/or VI. This is due to the formation of blood clots in the cavernous sinus which is often caused by infection by gram-positive bacteria It spreads contralaterally so that bilateral symptoms often occur(Olse et al, 2017).

Ophthalmoplegic pain that responds well to corticosteroid therapy includes carotid cavernous fistula, sarcoidosis, lymphoma and ophthalmoplegic migraine. Sarcoidosis and lymphoma often cause systemic symptoms. Aneurysms and meningiomas rarely cause

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orbital pain. Meningiomas do not respond well to corticosteroid therapy. Blood vessel abnormalities such as fistulascavernosaccharotid, migraineophthalmoplegiaand the aneurysm has no connection with a mass in the cavernous sinus or orbital apex as in STH.(Samatra et al, 2016).

Orbital pain is not a symptom of cranial nerve III palsy so this diagnosis is unlikely. Orbital cellulitis is the result of a bacterial infection that occurs in the bone. Usually occurs due to previous trauma or spread of the ethmoidal sinus. Clinical features include proptosis, eyelid swelling, conjunctival chemosis, and limited ocular motility. Cavernous sinus thrombosis can also be a cause of orbital cellulitis (Olse et al., 2017.

The patient in this case received methylprednisolone injection therapy 125 mg/ 8 hours and responded well to reduced pain, ptosis, proptosis and paresthesia as well as ophthalmoplegia. Administration of systemic corticosteroids to STH for 48 hours results in rapid improvement in clinical symptoms because of its anti-inflammatory properties so it can help rule out differential diagnoses although it cannot prevent recurrence.(Samatra et al, 2016) The period of administration is 2-4 weeks and then tapering off gradually over 2-6 weeks. (Lasam et al., 2016)The case report of Simanungkalit et al stated that pain in STH improved within 24-72 hours with intravenous dexamethasone at a dose of 20 mg/day and improvement in ptosis was also seen. If when you are discharged, eye movement is still limited, oral corticosteroids can be given at a dose of 1 mg/kg/day, then reduced gradually and continued according to the patient's clinical response (Olsen et al., 2017)Some cases that respond well to corticosteroid administration have a good prognosis (Filho et al., 2018).

## CONCLUSION

A male patient with this case must undergo a laboratory examination to be able to establish a related diagnosis, the results of the examination are normal, on the MRI examination there is thickening of the lateral rectus muscle. The patient was given methylprednisolone injection 125 mg/8 and responded well. The diagnosis of Tolosa Hunt syndrome in this case was confirmedbased on the presence of unilateral headache in the periorbital area accompanied by weakness in the nerves that move the eyeball (cranial nerves III and VI left), burning sensation and electric shock.in areaunilateral facial involvement indicating involvement of the V1 nerve.Chronic inflammation of the septa of the cavernous sinus wall in dense connective tissue can exert pressure that can penetrate cranial nerves III, IV, VI and V branches 1. Administration of systemic corticosteroids for 48 hours results in improvement of clinical symptoms.

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