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# Chronic Granulomatous Tolosa-Hunt Syndrome



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D. P. Samatra,\* I. M. P. Kesanda, and I. A. A. Indrayani

## ABSTRACT

**Background:** Tolosa-Hunt syndrome is a rare case, characterized by tenderness, persistent around the affected eye and ophthalmoplegia/paresis caused by granulomatous inflammation in the cavernous sinus region, supra orbital or orbital fissure. Although spontaneous remission may occur, even corticosteroid therapy has a very satisfactory effect. However, relapse can occur after remission. We report a case of granulomatous Tolosa-Hunt syndrome in women aged 47 years who suffer from recurrent Tolosa-Hunt syndrome attacks for 4 years on his left eye, there was a significant recovery after receiving steroid therapy.

**Case:** We report A 47 years old with recurrent pain in the left eye since 4 years, pain episode duration of 1-2 weeks, accompanied by double vision when having long or short distance viewing, and when climbing stairs. The patient left eye was protruded with blurred vision and difficulty in distinguishing green color. Left eye examination vision 1/300, green color discromatopsia, normal funduscopy, ptosis,

with paresis eye movement toward the superior, inferior, nasal and temporal. C-reactive protein and erythrocyte sedimentation rate were slightly elevated. ANA test was positive. In visual evoked potential, it showed latency elongation of the left face. Head MRI with contrast showed a isointense protrusion on the left cavernous sinus in axial cuts in T1 and T2. Head MRI T1 with contrast on coronal, axial cuts showed the appearance of convex lesions around the left cavernous sinus that enhanced with contrast.

**Conclusions:** The result was clinically and radiographically diagnosed as Tolosa-Hunt Syndrome (THS). Therefore, 10 mg dexamethasone therapy, 4 times a day for 3 days was lowered to three times on day 4, 2 times on the fifth day and one time at day 6. The patient showed clinical improvement. The patient continued 48 mg oral methylprednisolone therapy up to 3 weeks which then gradually decreased and planned head MRI 3 months later.

**Keywords:** Tolosa-Hunt syndrome, ophthalmoplegia and pain around the eyes

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## INTRODUCTION

Ophthalmoplegia or ophthalmoparesis followed by the retro-orbital or periorbital pain is quite common. Trauma to the blood vessels caused by internal carotid artery or cerebral aneurysm, carotid-cavernous fistula, cavernous sinus syndrome, tumors, inflammation or infection in sinusitis, periostitis, aspergillosis, mucormycosis, sarcoidosis, Wegener's granulomatosis, diabetic neuritis, horton's disease, ophthalmoplegic migraine and tolosa-hunt syndrome (THS).<sup>1</sup>

THS is one of the causes of ophthalmoplegia or ophthalmoparesis that accompanied by the retro-orbital or periorbital pain due to non specific inflammation (granulomatous or non granulomatous) in the cavernous sinus, supra orbital fissure or in the orbit. Tolosa was first found on a group of patients in 1954, the present diagnosis criteria have been revised by the International Headache Society. In which these criteria consist of, one or more episodes of pain in unilateral orbital area that persisted for several weeks or longer if not treated, followed by paresis of one or more of the third, fourth, and sixth cranial nerves, can be accompanied

by paresis of the first branch of the fifth cranial nerve (ophthalmic nerve) in rare cases may be accompanied by disorders of the optic nerve and can be accompanied by proptosis. On magnetic resonance imaging (MRI) or biopsy examination showed nonspecific inflammation appearance. Pain and paresis will improve within 72 hours with corticosteroid treatment, after removing the cause elsewhere.<sup>2</sup>

In this paper, we reported a chronic granulomatous tolosa - hunt syndrome case.

## CASE REPORT

A 47 years old woman was treated to Sanglah Central General Hospital (RSUP), with recurrent pain in the left eye since 4 years, pain episode duration of 1-2 weeks, accompanied by double vision when having long or short distance viewing, and when climbing stairs, the patient left eye was protruded with blurred vision and difficulty in distinguishing green color. There is no history of fever, infection around the paranasal sinuses or head injury. There is no history of similar illness

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in the family, headache, and migraine. The patient only took pain killer medication to reduce pain in the left eye.

Left eye examination vision 1/300, green color discromatopsia, normal fundusopic, ptosis, with paresis eye movement toward the superior, inferior, nasal and temporal. Normal direct light reflex and slow indirect light reflex. In the right eye, the pupil is normal, normal direct light reflexes and slow indirect light reflexes. Proptosis on the left eye. Sensation in the ophthalmic branch innervation is still good, direct and indirect corneal reflex directly on the right and left eye is normal. There was no other systemic and neurological signs or symptoms found.

The complete blood count, liver and kidney function test, blood sugar examination, and electrolytes examination were within normal limits. C-reactive protein and erythrocyte sedimentation rate were slightly elevated. ANA test was positive. In the cerebrospinal fluid analysis that included microscopic examination, biochemical, bacterial and viral culture were within normal limits. In visual evoked potential, it showed latency elongation of

the left face, but the electromyogram was within normal limits. In head MRI with contrast, there was a isointense protrusion on the left cavernous sinus in axial cuts in T1 and T2 - weighted image, indicated by white arrows in [figure 1](#). These lesions were enhanced in contrast injection, which indicated by the black arrow in [Figure 2](#). Left internal Carotid Artery and willis circle were still intact. There was no tumor-suspected mass.

The result was clinically and radiographically tended toward the diagnosis of THS. Therefore, 10 mg dexamethasone therapy, 4 times a day for 3 days was lowered to three times on day 4, 2 times on the fifth day and one time at day 6. The patient showed clinical improvement, which included reduced the left eye pain, improved proptosis, improved ptosis, improved diplopia complaints and movable left eye in all directions, thus improving visual acuity 1/60 and green color discromatopsia disappeared. The patient had continued 48 mg oral methylprednisolone therapy up to 3 weeks which then gradually decreased and planned for the control head MRI 3 months later.

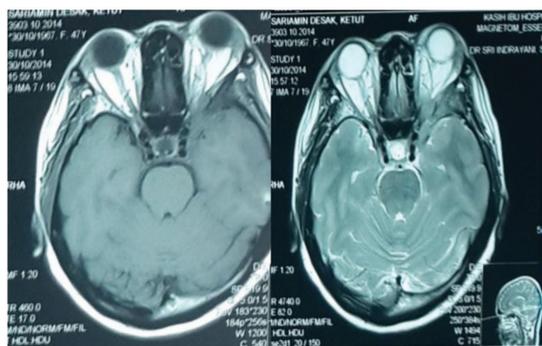
## DISCUSSION

THS as a result of non-specific inflammation, causing suppression, in the area around the cavernous sinus, although to date the mechanism or the etiology of this disease has not clear. This process can lead to an emphasis on structure within the cavernous sinus (cranial nerves III, IV, V1, VI and internal carotid artery) and pain around the eye ball.<sup>3</sup>

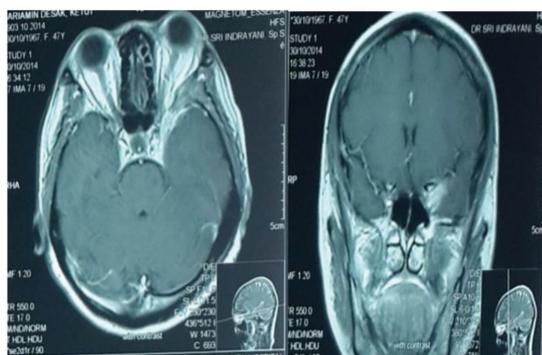
THS is usually reported unilateral, with no predisposition between right or left sinus cavernous, THS only happens about 5% bilaterally. THS occurs between the ages of 5-75 years and occurs equally in men and women. THS most often affects nerves III, VI, the first branch of nerve V, then nerve IV.<sup>4,5</sup>

THS diagnosis requires exclusion of other ophthalmoplegia or paresis causes with pain such as infection, malignancy or vascular lesions based on clinical examination, laboratory and neuroradiology. Due to the location of the lesion and its small size around the cavernous sinus making it difficult and dangerous for biopsy, imaging plays an important role in diagnosing this disease. In case where biopsy can be done, it is found a nonspecific granulation tissue in the cavernous sinus, pachymeningitis in the supraorbital fissure and necrotizing inflammation within intracavernous and a part of intracranial internal carotid in pathology examination.

In the radiological evaluation before by using plain x-ray, orbital Venography, cerebral angiography and computed tomography (CT) head scan has been more widely used for the diagnosis of THS



**Figure 1** Head MRI with T1 and T2 Axial cut showed the appearance of convex lesions around the left cavernous sinus.



**Figure 2** Head MRI T1 with contrast on coronal, axial cuts showed the appearance of convex lesions around the left cavernous sinus that enhanced with contrast.

with unsatisfactory results. After the era of MRI, this method is considered the best in diagnosing THS. Where on MRI, there are convex curved isointense enlargement in the cavernous sinus on T1-weighted images and isointense or slightly hypointense on T2-weighted images. These lesions are enhanced with contrast administration.

However, the MRI result was not specific to THS because lesions such as meningioma, lymphoma, and sarcoidosis can have an appearance of THS and THS can have a normal MRI result. In our patients MRI showed lesions on T1 and T2 isointense around the left cavernous sinus. The lesion was enhanced with contrast administration.

These lesions lead to inflammatory granulomatous processes, supported by increased inflammatory markers. In this case, inflammatory granulomatous cause a sharp decline in vision and inflammation of the soft tissue around the eye, resulting in protrusion of the eyeball. In this case a biopsy was not done because the procedure was invasive and based on clinical examination, laboratory and radiological investigation leads to the inflammatory process. In 2006, DK et al Gyun, reported a cases of THS on a 11 years old girl, with a clinical manifestation of ophthalmoparesis with pain in the left eye movement accompanied with proptosis on the left eye. By the non contrast head MRI, T1 and T2, it was discovered an enhanced isointense biconvex lesion with contrast administration, it was found also an increased inflammation marker so that biopsy was not performed. Patients were given steroid therapy and found to have significant improvement after 3 weeks, MRI is repeated 3 months later and granulomatous lesions was not found again.<sup>3,7,8</sup>

Although spontaneous improvement may occur in THS case, but treatment with corticosteroids may decrease pain within 24 - 48 hours, and improve the cranial nerve paresis in 2 weeks. The recommended steroid therapy for THS is, with prednisone 1-1.5 mg/kg/day, for optimal dosage, duration and alternative therapy was still only a little information available. In these patient's dexamethasone injection is used as steroid therapy and showed satisfactory results obtained in which complaints and clinical improvement is achieved within 2 days of therapy. Therefore, dexamethasone therapy is continued until day 6 and continued using methylprednisolone by considering the lighter side effects compared to prednisone for 3 weeks, after which it is planned gradually decrease steroid therapy and patient is planned to re MRI 3 months later to see the results of therapy.<sup>1,9,10</sup>

THS has a good prognosis, but 30-40% of THS cases patients who have been treated will have a relapse. It usually occurs on the same side, but can also occur on the contralateral side. Our patients

have experienced repeated attacks on the same side, although not previously been treated for steroid. This case is a unique case because it is a variant of rare chronic granulomatous cases by folding of the optic nerve and causing proptosis of the left eye.<sup>1</sup>

## SUMMARY

Tolosa-Hunt syndrome caused pain that is accompanied by ophthalmoplegia or paresis of the eye movement, the diagnosis is established by ruling out the other causes. One variant of this disease is the inflammatory granulomatous which can be accompanied by the optic nerve disorders and proptosis. In rare cases, the disease can be chronic. This disease generally responds well to corticosteroid therapy, but recurrence may occur in 30-40% of cases.

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