CASE REPORT

A Case Report: Tolosa Hunt Syndrome Presenting With Painful Trigeminal Neuropathy

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ABSTRACT

Tolosa-Hunt Syndrome (THS) is a rare idiopathic granulomatous inflammation of the cavernous sinus or superior orbital fissure, leading to unilateral periorbital pain and cranial nerve palsy. It can also cause neuropathic pain in the trigeminal nerve distribution, particularly in the ophthalmic (V1) and maxillary (V2) branches. We report a case of a 68-year-old woman presenting right eye ptosis, medial rectus weakness, right periorbital pain with electric-like quality especially when moving the eyeball and allodynia in the right eye area for the past two months. Contrast-enhanced brain MRI revealed right cavernous sinus thickening. Treatment with Prednisone 60 mg/day for 1 week and Gabapentin 300 mg/day for 1 month resulted in marked improvement of ptosis and pain.
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Keywords: Tolosa-Hunt syndrome, Trigeminal neuropathy, Painful Ophthalmoplegia, Unilateral Periorbital Pain, Idiopathic Inflammatory Syndrome

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INTRODUCTION

Tolosa-Hunt Syndrome (THS) is defined as one-sided orbital or periorbital pain associated with partial paralysis or weakness of either oculomotor, trochlear and/or abducens cranial nerves due to granulomatous inflammation in the cavernous sinus, superior orbital fissure or orbit and the ophthalmic branch of the trigeminal nerve might be affected. THS is classified as a rare disease, according to the National Organization for Rare Disorders (NORD), with characteristics of severe unilateral pain around the orbita accompanied by paralysis of the eye muscles. Visual Analog Scale (VAS) score is often used to assess pain levels in THS patients. This scale helps clinicians assess the severity of pain over time and guide treatment decisions. The prevalence of THS is 1 – 2 cases per million per year. There is no gender predilection among adults with THS, but it can occur at any age from the first to eighth decades of life. Until now, the exact etiology of THS remains unknown. Some theories suggest that THS is related to non-specific (non-granulomatous/granulomatous) inflammation in the cavernous sinus area and/or fissures due to excess fibroblast formation and percolation of the septa and walls of the cavernous sinus with lymphocytes and plasma cells. Persistent pain is often the initial symptom of this syndrome(1). Symptoms of THS are described in the International Classification of Headache Disorders (ICHD 3) as unilateral orbital pain associated with paresis of one or more of the third, fourth, and/or sixth cranial nerves (2).

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A 68-year-old woman presented to the neurology clinic of Dr. Soetomo General Hospital, Surabaya, Indonesia, with a two-months history of right eyelid drooping and periorbital pain prior to hospital admission. Initially, the patient experienced a dull, intermittent, mild pain in the right periorbital region. However, after two months the pain gradually intensified into a constant, sharp, electric-like pain with VAS of 6 particularly aggravated by eye movement. The pain was followed by the development of ptosis, prompting the patient to seek medical attention. The patient had no history of trauma, hypertension, diabetes mellitus, or stroke. Neurological examination revealed right eye ptosis (see Figure 1), decreased pupillary light reflex of the right eye, paresis of the medial rectus muscle, and allodynia according to the

dermatome of the ophthalmic branch of the trigeminal nerve. Contrast-enhanced brain Magnetic resonance imaging (MRI) showed thickening of the right cavernous sinus structure (see Figure 2). Other investigations were not conducted due to patient's refusal. The patient was treated with prednisone 60 mg/day for one week, which resulted in improvement ptosis, medial rectus function, and reduction of pain to a VAS score of 2. Allodynia symptoms were improved by administration of gabapentin 300 mg/day for one month. Written consent has been obtained from the patient prior to the publication of this manuscript.

DISCUSSION

THS is a rare neuro-immunological disorder affecting 1 – 2 per million people, characterized by the classic triad of unilateral orbital pain, cranial nerve paresis, and granulomas on MRI or biopsy(1). THS is diagnosed by criteria in ICHD-3, which includes unilateral headache in the orbit or periorbita and paresis of one or more of the ipsilateral third, fourth, and sixth nerves. Trigeminal

Fig 1: Clinical Condition of Ptosis

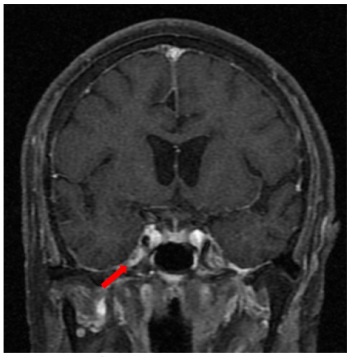

Fig 2: MRI Brain with Contrast

neuralgia (TN) may also occur in patients with THS. The ophthalmic (V1) and the maxillary (V2) branches of the trigeminal nerve enter the cavernous sinus through Meckel's cave and course along the lateral wall of the cavernous sinus. Therefore, inflammation within the cavernous sinus can lead to neuropathic pain in the distribution of the trigeminal(3). Although the patient's pain had an electric-like quality and was localized to the ophthalmic (V1) distribution—features suggestive of trigeminal neuralgia—other essential criteria from the ICHD-3 for classical TN were not met (Table 1). Notably, the pain was constant rather than paroxysmal, neurological deficits were present, and MRI demonstrated cavernous sinus inflammation consistent with THS. Therefore, the facial pain is better categorized as trigeminal neuropathy due to THS rather than classical TN. The patient's MRI revealed thickening of right cavernous sinus. MRI is the most valuable modality for differentiating THS from other THS-like entities with a sensitivity of approximately 80-90% and specificity of 70-85%. THS typically appears as localized, well-defined enhancement of the cavernous sinus or superior orbital fissure, often without the involvement of surrounding structures. In contrast, Cavernous Sinus Thrombosis due to infection presents as diffuse, poorly defined cavernous sinus enlargement, often accompanied by engorgement of the superior ophthalmic vein and spread of infection. Idiopathic Orbital Inflammation (IOI) often involves the orbit itself, appearing as a well-defined orbital mass, with extraocular muscle, or fat involvement, showing significant gadolinium enhancement but without cavernous sinus involement. Meningiomas typically presents as well-defined extra-axial masses with homogeneous enhancement, often attached to the dura, whereas metastases typically present as multifocal, irregularly enhancing lesions. Vasculitis, such as Giant Cell Arteritis or Sarcoidosis, may show vessel wall enhancement or granulomatous masses, with vasculitis potentially affecting multiple sites, including the cavernous sinus. Therefore, MRI findings—specifically the pattern of enhancement and anatomical involvement—are essential for distinguishing THS from other conditions. Although THS can resolve spontaneously, corticosteroids remain the treatment of choice due to their high response rate. Common regimens include high-dose steroids (>0.5–1 mg/kg/day), tapered over 3–4 months, though optimal dose and duration remain debated. In our case, one week administration of prednisone led to the improvement of ptosis and pain, followed by a one-month course of gabapentin to relieve allodynia. Short course steroid therapy was implemented to prevent the long-term adverse effects of systemic steroids. The majority of patients experience significant improvement or complete resolution of pain and ophthalmoplegia within 2-3 weeks of initiating corticosteroids, particularly with high-dose oral prednisone. However, the recurrence rate of THS is notable, with around 20-30% of patients experiencing a relapse, often within the first year after treatment(2). Corticosteroids that can be given are prednisone, methylprednisolone, and dexamethasone orally or intravenously(1). Administration of methylprednisolone 40 mg/day for 7 days can eliminate pain complaints in 60-year-old men with comorbid diabetes mellitus but does not restore visual disturbances and ptosis(4). In another case, administration of dexamethasone 80 mg/day for 1 month can reduce oculomotor disorders dramatically, and a repeat MRI after 3 months of administration showed that the lesions in the cavernous sinus had disappeared(5). Administration of gabapentin 1200 mg/day can help reduce pain symptoms in patients(3). THS is a very rare case, especially in this patient, who also has trigeminal neuropathy. Despite the rarity of this condition, it is crucial for clinicians,

particularly in neurology, to be vigilant and not neglect the diagnosis of THS in patients presenting with periorbital pain so that the diagnosis of THS can be detected and treated appropriately. THS if left untreated or misdiagnosed, complications related to prolonged nerve damage or the underlying inflammatory process might arise which will worsen the patient's quality of life.

CONCLUSION

THS is a rare but treatable cause of painful ophthalmoplegia. MRI is critical for diagnosis and for distinguishing THS from other orbital and cavernous sinus pathologies. Corticosteroid therapy typically results in rapid symptom relief, and adjunctive use of gabapentin may benefit patients experiencing concurrent trigeminal neuropathy. Although this condition is rare, it is essential for clinicians to remain attentive and not overlook the diagnosis of THS in patients presenting with periorbital pain, ensuring that THS is identified and managed appropriately.

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