

Case Report

Tolosa-Hunt Syndrome: A rare cause for an isolated ocular nerve palsy in an elderly male

S Pirasath¹, T Kumanan², M Malaravan³¹Registrar, Professorial medical unit, Teaching Hospital, Jaffna, Sri Lanka²Consultant Physician and Senior Lecturer, Professorial medical unit, Teaching Hospital, Jaffna, Sri Lanka³Consultant ophthalmologist, Teaching Hospital, Jaffna, Sri Lanka***Corresponding Author:**

S Pirasath

Email: selladuraipirasath81@gmail.com

Abstract: Tolosa-Hunt syndrome (THS) is essentially a clinical diagnosis of exclusion; painful ophthalmoplegia caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure. We reported a 70 year old male with THS who responded to corticosteroids dramatically and made an uneventful complete clinical recovery.

Keywords: tolosa-hunt syndrome, ocular nerve, corticosteroids.

INTRODUCTION

Tolosa-Hunt syndrome (THS) is rare disorder indicated by recurrent painful ophthalmoplegia caused by non-specific inflammation of the cavernous sinus or superior orbital fissure. It leads to palsies of the third, fourth or sixth cranial nerves and first and second divisions of the trigeminal nerve. Prompt clinical response to steroids is the hallmark of this condition. The clinical presentation of THS has a wide differential diagnosis, and timely and appropriate imaging – as an adjunct to pertinent laboratory investigations – can greatly assist clinicians with early accurate diagnosis and management. We reported a 70 year old male with THS who responded to corticosteroids dramatically and made an uneventful complete clinical recovery.

CASE REPORT

A 70 year old male presented with a history of right sided headache and drooping of right eyelid lasting for a period of 14 days. He also complained impairment of upward and inward movements of right eye. He had a past history of stable angina (Class III) and underwent primary coronary angioplasty 12 years back. On examination, his vital signs were normal. Examination of the cranial nerves revealed right side complete ptosis and impairment of adduction and elevation of the right eye, consistent with an oculomotor (IIIrd) nerve palsy. The right pupil was mildly dilated but responsive to light. No sensory or motor loss was detected in face. Remaining cranial nerves examinations were normal. His ocular examination (Anterior and Posterior segments) findings were normal. Blood work and lumbar puncture (LP) were non-specific. The white cell count (WCC) was mildly elevated and the

erythrocyte sedimentation rate (ESR) and C reactive protein were significantly increased. His retroviral screening was negative. The further laboratory tests for antineutrophil cytoplasmic antibody (c-ANCA), antinuclear antibody (ANA), anti-double-stranded DNA (anti-dsDNA) were negative. A magnetic resonance imaging (MRI) scan of brain revealed the abnormal area in the cavernous sinus of intermediate intensity on T1W1. This is consistent with the pathological process of THS that is granulomatous inflammation. In the appropriate clinical setting of painful ophthalmoplegia and MR findings of a cavernous sinus abnormality suggests the diagnosis of THS. The patient was treated with oral steroids. His symptoms were improved dramatically over some days. He had complete resolution of the ptosis as well as the ocular movements within three months. He has no recurrence of symptoms during one year follow-up.

DISCUSSION

Tolosa-Hunt syndrome (THS) is characterized by recurrent painful ophthalmoplegia [1] caused by non-specific inflammation of the cavernous sinus or superior orbital fissure [2] (SOF). It involves palsies of the third, fourth or sixth cranial nerves and first and second divisions of the trigeminal nerve [3]. Our patient presented with isolated third nerve palsy. First case was described as granulomatous periarteritis of cavernous carotid in 1954 [4]. Six cases with similar clinical findings were described in 1961 [5]. The low-grade non-specific inflammation of the cavernous sinus was proposed as the cause of the syndrome [5]. The proliferation and infiltration of fibroblasts were noted in wall of cavernous sinus with lymphocytes and

plasma cells [6]. The International Headache Society (IHS) has proposed the diagnostic criteria for THS in 1998 and has further revised in 2004 [7]. The MRI is a crucial role in a patient presenting with features of THS and helps to exclude the differential diagnosis of steroid responsive painful ophthalmoplegia [8, 9]. Administration of systemic steroids produces a dramatic response in a patient with THS [10]. However sarcoidosis and lymphoma will often have systemic symptoms and meningiomas will not resolve with steroid therapy. THS essentially remains a diagnosis of exclusion. Distinctive MRI findings and rapid resolution of clinical symptoms with steroid therapy are characteristic. It allows differentiating THS from other conditions of painful ophthalmoplegia [11, 12]. The prompt clinical response to steroid therapy, unremarkable laboratory work up for other medical comorbidities and characteristic nonspecific inflammation in MRI study confirm the diagnosis in this patient.

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