ABSTRACT

Tolosa-Hunt syndrome (THS) is a rare benign treatable cause of painful ophthalmoplegia with frequent VI, III, IV cranial nerve (CN) involvement. Optic and trigeminal nerve involvement is rare but described. The disease is characterized by severe preceding or concomitant headache and remarkable response to steroids. Although labelled benign residual neurology is not uncommon in THS.

We present a case of THS in a middle-aged Sri Lankan man who presented initially with painful vertical diplopia with evidence of isolated right IV CN involvement. Contrast enhanced CT (CECT) scan was normal except for sinusitis. He was treated for idiopathic IV nerve palsy and sinusitis. Within the course of a month, he developed severe episodic ipsilateral headache with autonomic symptoms in clusters with right sided ptosis, progressive visual impairment manifesting as disappearance of diplopia, with normal fundoscopy, normal inflammatory and infective markers and autoimmune profile. Repeat CECT and MRI brain showed enhancing lesion of cavernous sinus extending to orbital apex consistent with THS. Biopsy was not performed due to lack of feasibility and procedural risk. He showed dramatic improvement with steroids with re-appearance of diplopia and complete resolution of neurology at the end of 4 weeks.

THS should be considered in a patient presenting with disappearing painfuldiplopia. Headache may mimic primary headache syndromes and may not always occur preceding ophthalmoplegia. CECT is may be normal at outset and could be repeated but MRI is mandatory and biopsy carries high risk. Steroid responsiveness can be diagnostic if more sinister differentials are ruled out.