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Tolosa-Hunt syndrome following recurrent facial palsies

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Abstract

Tolosa-Hunt syndrome is typically associated with an inflammatory lesion in the cavernous sinus or orbital fissure, often requiring steroids for symptom resolution. In this report, we describe a case of Tolosa-Hunt syndrome preceded by several years' history of idiopathic recurrent facial palsies. The spontaneous resolution of THS in our case as well as prior facial nerve involvement supports the hypothesis that Tolosa-Hunt syndrome is part of a spectrum of idiopathic recurrent cranial neuropathy.

INTRODUCTION

The Tolosa-Hunt syndrome (THS) has classically been described as an episodic orbital pain associated with paralysis of one or more of the 3rd, 4th and 6th cranial nerves, resolving spontaneously or with the commencement of steroids. Often the episode is associated with a granulomatous lesion in the cavernous sinus or orbital fissure.[1] Over the years, there have been cases reporting the involvement of cranial nerves outside the cavernous sinus.[2-5] This raises the possibility that the syndrome is in fact part of a larger spectrum of idiopathic recurrent cranial neuropathy.[4]

In this article, we report a case of THS which was preceded by several years' history of recurrent bilateral facial nerve palsies.

CASE REPORT

A 37 year old female presented in April 2009 with a 5 day history of left retro orbital sharp, stabbing pain and diplopia. On further enquiry, her neurological history dated back to the year 2000 when she first developed a right sided facial weakness. At the time, she was managed as having Bell's palsy and treated with a course of acyclovir and steroids. Her symptoms and signs resolved after 10 days and she was left with intermittent stabbing pain over the right side of her face. She re-presented in 2002 with a recurrence of the right facial weakness. Blood investigations were normal including ESR. Nerve conduction studies done at the time showed a prolonged distal motor latency of the facial nerve. Magnetic resonance imaging (MRI) with gadolinium of the brain was normal. She was again treated with a tapering dose of oral steroids for a presumed diagnosis of recurrent Bell's palsy. Her facial weakness gradually resolved. Six months later, she developed a third episode of facial weakness, this time affecting the left facial nerve. The episode was associated with sharp, stabbing pain. Oral steroids were started. Blood investigations including serum ACE and cerebrospinal fluid (CSF) analysis were unremarkable. MRI brain was also normal. High resolution CT chest, looking for evidence of sarcoidosis was done which was normal. Despite completing a course of steroids, she was left with a residual mild left facial weakness and intermittent left hemifacial spasm. She remained stable until the current episode, seven years later.

Her current examination revealed a left 6th cranial nerve palsy. There was also a mild left facial weakness, residual from her previous attack. The rest of her cranial nerve examination was normal, including pupillary examination and limb examination. Blood investigations including autoantibody screen (ANA, ANCA, antiphospholipid, lupus anticoagulant), Lyme serology, ESR were within normal limits. CSF analyses were as follows: red blood cells 630, leucocytes 3, glucose 3.6, IgG 0.11 (0.01-0.03), serum IgG 1300 (931-1916), albumin 0.41 (0.1-0.25), serum albumin 40 (35-50), IgG/Alb 27% (<12), oligoclonal bands were not detected. Tuberculosis PCR and culture was negative. An MRI brain with gadolinium showed an inflammatory lesion of the left orbital apex with subtle extension into the left cavernous sinus (Figure 1). MR angiography of the cerebral vessels and formal cerebral angiography were normal. The patient opted not to have any steroid treatment

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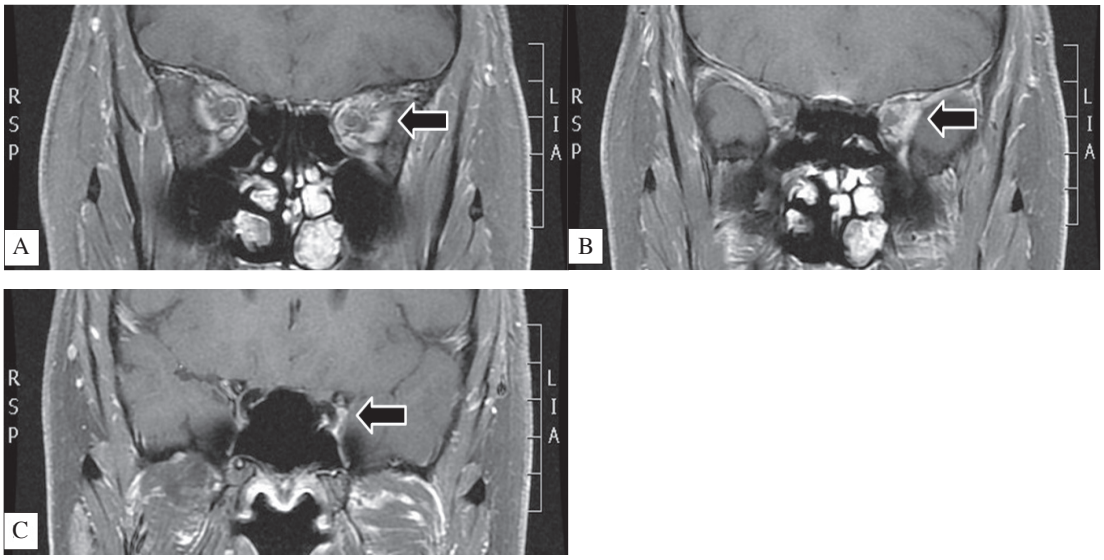

Figure 1. MRI at initial presentation with left 6th cranial nerve palsy. (A) and (B): MR coronal T1 weighted imaging post gadolinium with fat suppression. The arrows highlight an enhancing lesion in the perioptic and tendinous insertion of the left lateral rectus causing a fullness of the left orbital apex. (C): The arrow highlights the enhancing lesion extending to the left cavernous sinus.

on this occasion and her symptoms resolved spontaneously within two weeks. A repeat MRI of the brain with gadolinium was done 3 months later showing improvement of the enhancing lesions (Figure 2).

DISCUSSION

We describe a case of recurrent cranial neuropathy affecting the 6th and 7th cranial nerves. The most recent neurological presentation of painful left 6th cranial nerve palsy fulfilled the diagnostic criteria of THS and included the localisation of an enhancing lesion in the orbital apex. However, the involvement of both facial nerves, many years prior to the current episode is atypical for THS and suggests a more generalised inflammatory process.

In 1954, Eduardo Tolosa described a patient with painful left opthalmoplegia, visual loss and reduced sensation over the ophthalmic division of the trigeminal nerve, associated with a granulomatous inflammation of the cavernous sinus and carotid artery.[6] Hunt *et al*, in 1961, identified six patients with a similar presentation. They found the aetiology to be less well defined but postulated probable inflammation within the cavernous sinus as well as documenting a prompt response to steroids in two of their patients.[1] The term "Tolosa-Hunt syndrome" was suggested by Smith *et al* in 1966 who emphasised the use of steroids as a diagnostic tool in their patients.[7] Many similar cases have since been reported. The involvement of the facial nerve along with features of THS has also been described suggesting an

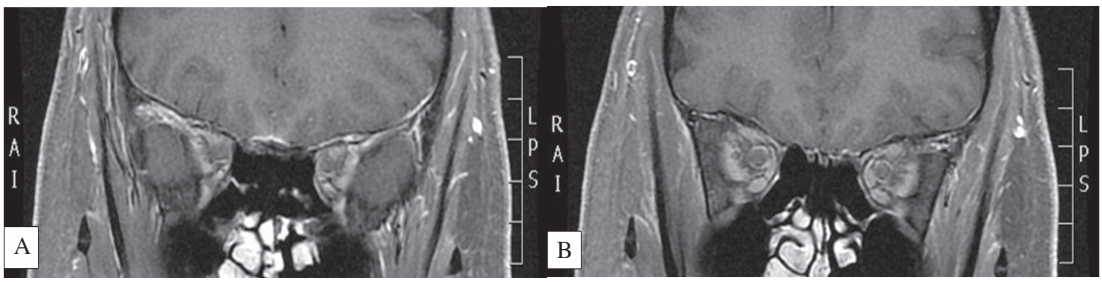

Figure 2. MRI performed 3 months following spontaneous resolution of symptoms. (A) and (B): MR coronal T1 weighted imaging post gadolinium with fat suppression demonstrates an improvement of the enhancing lesion in the left orbital apex.

extension of the area of inflammation to outside the cavernous sinus.[2-5,8,9] Miwa *et al* however, documented facial palsy preceding THS by up to three years in three of their patients.[4]

In the current case, we describe the occurrence of recurrent facial palsy preceding the features of THS. MRI brain scan with gadolinium done during the facial palsy episodes failed to show any abnormality but repeat MRI with gadolinium during the episode of painful left 6th nerve palsy showed a corresponding small enhancing lesion in the lateral aspect of the left orbital apex. This concurs with the literature on neuroimaging in Tolosa-Hunt syndrome which typically shows an intermediate signal on T1 weighted MR imaging, enhancing with contrast.[10-13] The abnormal lesion typically reduces in size with corticosteroid therapy. In the case of our patient, a repeat MRI scan showed a regression in the size of the enhancing lesion without steroids. The presence of a raised CSF IgG index in our patient suggests an inflammatory process within the CNS but is non-specific as to the underlying aetiology. This finding, however, lends further support as to the inflammatory nature of recurrent cranial neuropathy. Ideally, a biopsy of the enhancing lesion would have helped to further establish the diagnosis. In THS, granulomatous changes with epitheloid cells and occasional white cells have been described.[14] As the symptoms in our patient were resolving spontaneously, the potential risks associated with a neurosurgical biopsy of the area in question were unacceptable.

There are three possibilities for the occurrence of recurrent cranial neuropathies in our patient. Firstly, the patient's recurrent cryptogenic isolated facial palsies may have been unrelated to the current Tolosa-Hunt presentation; secondly, the recurrent facial palsies and the recent presentation of painful partial ophthalmoplegia may be part of an as yet to be identified underlying condition; or thirdly, THS may in fact be part of an idiopathic recurrent cranial neuropathy spectrum of which painful ophthalmopegia and recurrent facial neuropathies can occur. Given the thorough investigations and long duration of the history, the first and second conclusions appear to be unlikely and an idiopathic recurrent cranial neuropathy spectrum is the most likely cause for our patient's recurrent symptoms.

We would encourage further publications on the atypical presentations of THS, including those involving cranial neuropathies outside the cavernous sinus as this will add to our understanding of this fascinating disorder.

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