Tolosa Hunt Syndrome: a rare cause of Hypopituitarism

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Introduction

Tolosa Hunt Syndrome (THS) is a idiopathic inflammatory granulomatous condition affecting the cavernous sinus and/or orbital apex causing painful ophthalmoplegia. It is a rare syndrome affecting 1 person per million per year. We present a rare case of THS resulting in hypopituitarism.

Background

Initially described by Tolosa[1] in 1954 and expanded upon by Hunt[2] in 1961 the condition is unilateral, but can be bilateral and is strongly associated with periorbital headaches and pain. Patients exhibit paralysis of the III, IV, or VI cranial nerves and are often noted to have proptosis and/or a dilated pupil.

The diagnosis of THS has to be made following clinical evaluation and neuroimaging. There are many other causes for compressive lesions in the cavernous sinus, notably aneurysm, fistula, neoplasia, thrombosis or lymphoma, that can cause the same clinical presentation which have to be ruled out prior to the the diagnosis of THS. The International Headache Society supplied a specific diagnostic criteria for THS (Table 1).

If left untreated THS usually spontaneously resolves after about two months but is normally highly responsive to glucocorticoids when given. It is considered a benign condition but rarely has been known to cause permanent neurological deficits and cause hypopituitarism.

Case

A 45-year-old female presented with a ten day history of headache, periorbital pain and diplopia. Her past medical history included bipolar disorder and bilateral below knee amputation from rail accident. She took mirtazepine, olanzapine and epilim for bipolar.

International headache Society THS diagnostic criteria
• Unilateral headache
• Granulomatous inflammation of cavernous sinus or orbit
• Paresis of one or more of III,IV or VI cranial nerves
• Symptoms not accounted better by another diagnosis

• Evidence of causation demonstrated by both:
  • Headache preceding ophthalmoplegia
  • Localised headache around ipsilateral brow and eye

Table 1.

On examination, a left abducens nerve palsy was noted with no other neurological or ophthalmological abnormalities. Haematology, biochemistry, autoimmune screen, angiotensin converting enzyme (ACE) were normal. MR angiography showed a hyper-intense lesion in the left of the cavernous sinus suggesting inflammatory tissue, but no vascular or pituitary abnormalities noted. (Fig. 1)

A diagnosis of THS was made, and prednisolone 60mg/day was commenced. Ophthalmic pain subsided within 48 hours; ophthalmoplegia resolved over 3 months leading to steroid cessation.

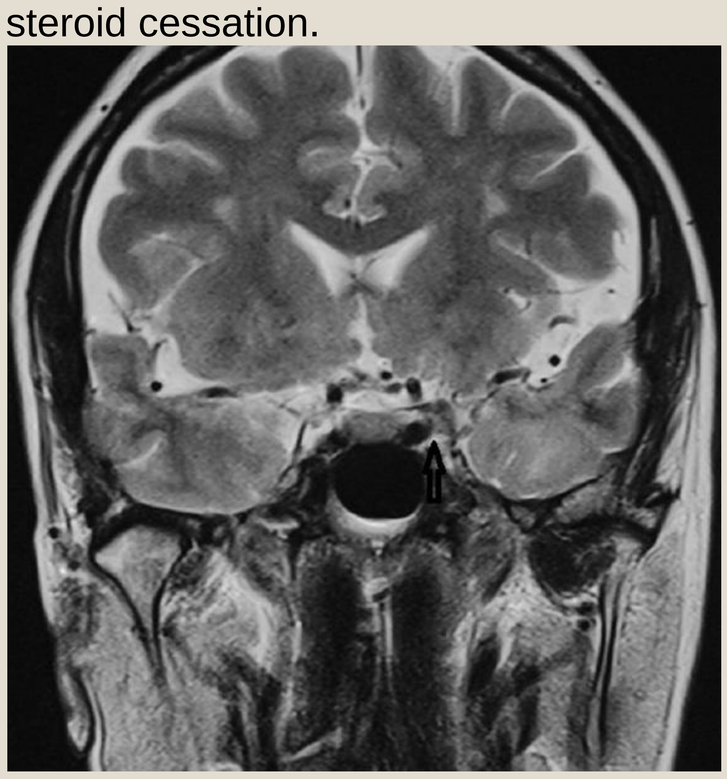

Fig.1. Tolosa–Hunt syndrome demonstrated by constructive interference steady state magnetic resonance imaging Wani NA[1], Jehangir M[1], Lone PA[2].

Progression

Six months later the patient developed polyuria, polydipsia, tiredness and weight loss. Biochemistry revealed new onset type 2 diabetes mellitus and secondary hypothyroidism. Symptoms improved with levothyroxine and diabetes treatment, but full pituitary evaluation was not undertaken. Evaluation 10 years later by an Endocrinologist revealed partial anterior hypopituitarism: secondary hypothyroidism, secondary hypogonadism and growth hormone deficiency. (Table 2.)

Test	Value	Reference range
TSH	1.4 miu/L	0.3-5.0 miu/L
FT4	6.9 pmol/L	9-25 pmol/L
LH	<0.5 iu/L	1.8-12 iu/L
FSH	0.8 iu/L	1.5-12.4 iu/L
IGF1	44 ug/L	88-141 ug/L
Short synacthen	normal	
Paired osmolalities	normal	

Table. 2 Pituitary Functions results

Discussion

THS is a rare cause of hypopituitarism with unknown aetiological mechanism. Fatalities with co-existing cerebral venous thrombosis have been reported. Partial hypopituitarism is common, but panhypopituitarism including Diabetes Insipidus is reported. Although high dose steroid treatment resulting in full pituitary function recovery is noted, residual chronic hypopituitarism is common.

Learning points

1. THS diagnosis should be suspected with presentation of painful ophthalmoplegia and full pituitary evaluation should be undertaken.
2. Symptoms usually resolve with high dose steroids but hypopituitarism is a recognised chronic complication.

References

1. Tolosa, E.. Periarteritic lesions of the carotid siphon with the clinical features of a carotid infraclinoidal aneurysm. J Neurol Neurosurg Psychiatry 1954; 17:300.
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