Bilateral visual loss in the Tolosa-Hunt syndrome

Perda visual bilateral na síndrome de Tolosa-Hunt

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SUMMARY

This paper describes a 65-year-old man with Tolosa-Hunt syndrome who presented with unilateral painful ophthalmoplegia and bilateral visual loss. Cranial CT scan revealed an enhancing mass at the right superior orbital fissure and optic canal but a biopsy showed only chronic inespecific inflammation. Treatment with corticosteroid resulted in marked improvement although one eye remained blind. The inflammatory process involved the right superior orbital fissure and optic nerve and presumably extended across the skull base to involve the contralateral optic nerve.

Key words: Tolosa-Hunt syndrome - Painful Ophthalmoplegia

INTRODUCTION

The Tolosa-Hunt syndrome (THS) is a non specific inflammatory condition of the cavernous sinus and superior orbital fissure area. Clinical findings include headache, involvement of the third, fourth and sixth cranial nerves and sensory loss in the ophthalmic division of the trigeminal nerve [1]. Severe visual loss is uncommon and usually restricted to the side of the ophthalmoplegia [2-7].

This paper documents one patient with THS who presented with the unique combination of unilateral painful ophthalmoplegia and bilateral visual loss.

CASE REPORT

A 65-year-old previously healthy man was admitted to the hospital because of a two months history of increasingly severe right sided headache radiating from the right retroorbital region and progressive visual loss in the right eye (OD). Two weeks prior to admission the headache worsened and he became blind in OD. One week later he noticed droopiness of the right eyelid and blurred vision in the inferior field of vision on the left eye (OS).

On examination, the vision was no light perception in OD and 20/20 in OS. There was an almost complete ptosis and absence of elevation of OD. Adduction, abduction and depression of this eye as well as the movements of the left eye were normal. There was no proptosis. Slit lamp examination and intraocular pressure measurements were normal in both eyes. The right pupil was slightly larger than the left and showed no direct reaction to light but would react normally when the left eye was illuminated. Ophthalmoscopy showed only slight paleness of the right optic disc. The left eye was normal. Goldmann perimetry disclosed an inferior depression of the isopters in OS.

General physical and neurological examination were otherwise unremarkable. The results of a complete blood count, erythrocyte sedimentation rate, glucose, VDRL, FTA-ABS, creatinine, serum protein electrophoresis, electrolites, PPD and chest X-ray were normal. A spinal tap was normal except for the

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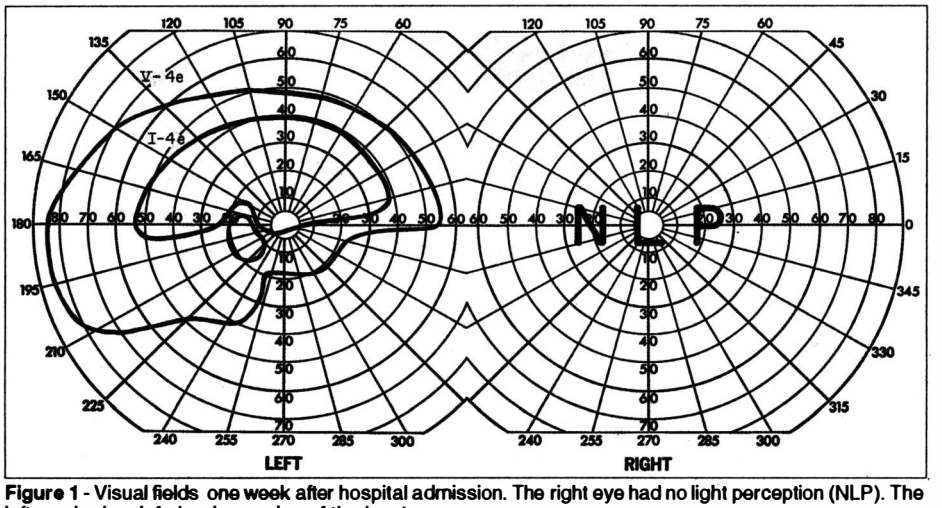

Figure 1 - Visual fields one week after hospital admission. The right eye had no light perception (NLP). The left eye had an inferior depression of the isopters.

finding of 11 cells, mostly lymphocytes. A cranial CT scan did not show abnormalities but only thick axial cuts were obtained. A right temporal artery biopsy was negative.

His headache persisted and there was progressive worsening of the ophthalmoplegia. One week later he had complete third and fourth nerve palsies. A repeat visual field showed worsening of the defect in OS (Figure 1). Treatment with 100 mg of oral prednisone a day was initiated. A few hours after the first dose the pain was gone. Over the following weeks there was gradual improvement of the ophthalmoplegia on the right side and of the field defect in OS. Prednisone was slowly tapered to 10 mg a day. Two months later there was only 1 mm of right ptosis. The right eye remained blind but the left visual field had returned to normal. The right optic disc showed severe atrophy and the left appeared normal.

Shortly thereafter, the headache recurred. A repeat CT scan revealed an enhancing mass occupying the right superior orbital fissure and enlargement of the optic nerve at the optic canal and orbital apex (Figures 2 and 3). A biopsy through a right frontotemporal craniotomy was performed in order to rule out an underlying neoplastic process. At surgery, the chiasm, optic nerves and adjacent structures seemed normal. The optic canal was unroofed but no tumor could be found. It was remarkable that there was only soft tissue separating the medial side of the optic canal and the sphenoid sinus. The impression was that the bone was decalcified in this area. Several biopsy specimens were obtained from the intracranial and intracanalicular portions of the right optic nerve, the adjacent dura and the bone of the optic canal.

Histologic sections showed only perivascular infiltration of lymphocytes in the optic nerve and dura mater. Special stains for fungi and acid fast bacteria were negative. The pathologic diagnosis was chronic inespecific inflammatory process, consistent with the diagnosis of THS.

Post operative recovery was uneventful. Follow up examinations up to four years later showed that his condition was unchanged. Headache had not recurred.

DISCUSSION

This case fulfils the criteria for the diagnosis of Tolosa-Hunt syndrome [1]. However, it showed the unusual finding of bilateral visual loss with complete optic atrophy in one eye. Visual loss is uncommon in the THS. Kline [1] reviewed 145 cases of the syndrome reported up to

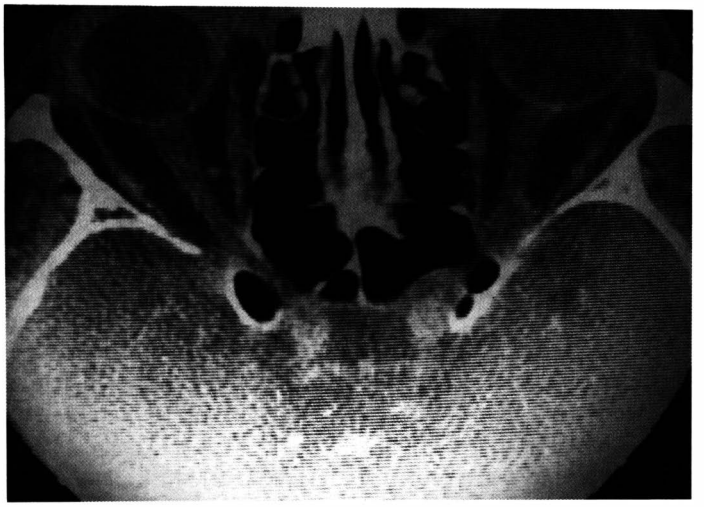

Figure 2 - CT scan with contrast showing enlargement of the right optic nerve at the orbital apex and optic canal.

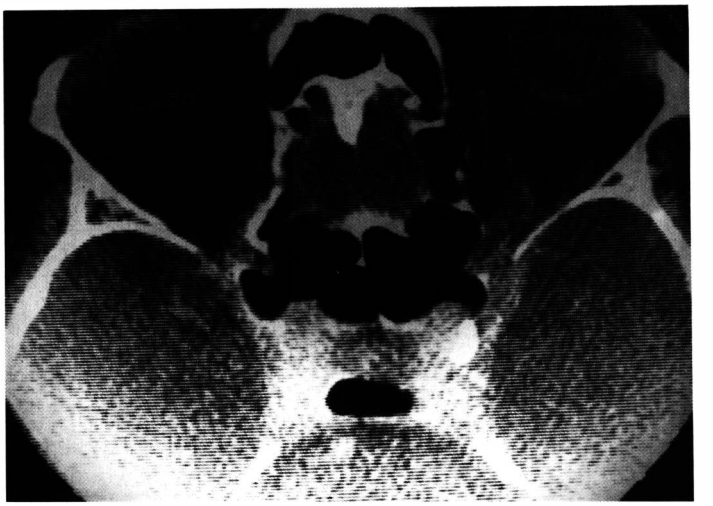

Figure 3 - Enhancing mass lesion at the right superior orbital fissure.

1982. Analysis of these patients revealed only 10 with documented visual loss [2-8]. The optic nerve involvement was always unilateral and generally improved with corticosteroid treatment although 4 patients developed optic atrophy [2,4,6,7].

The present case is unusual because of the complete optic atrophy and blindness that developed on the side of the ophthalmoplegia and the contralateral optic nerve involvement. This is remarkable since THS has generally been considered a unilateral disease. However, due to the rarity of detailed post mortem studies, very limited information on the extent of the pathologic process is available in this condition. Until recently, only Tolosa's original case had been studied at autopsy. A second, still unreported case was studied at the University of California San Francisco. A 75-year-old patient had painful ophthalmoplegia with involvement of the left II through VIII cranial nerves. Treatment with high dose of steroids was complicated with gastrointestinal bleeding that culminated with his death. At autopsy, there was thickenning of the dura overlying the left cavernous sinus, clivus and superior orbital fissure. These changes extended across the midline to involve the right side. Microscopic sections showed granulomatous inflammation containing histiocytes, lymphocytes and plasma cells, This case shows that the pathologic process in the THS may not be restricted to the cavernous sinus and superior orbital fissure. It can be much more diffuse, involving other cranial nerves and extending to the contralateral side. Six out of 146 cases reviewed by Kline were bilateral although none of them had bilateral optic nerve involvement [1].

In the present study, the CT scan and histologic findings indicated that the right sided blindness was caused by the extension of the inflammatory process to the right optic nerve at its intracranial, intracanalicular or proximal orbital segment. Presumably, the contralateral visual field loss was caused by extension of the inflammatory process across the skull base to involve the opposite optic nerve. Although the altitudinal type of field defect in OD suggested an ischemic event, its complete reversal with treatment is much more compatible with an inflammatory process.

The presence of a mass at the superior orbital fissure and the enlargement of the optic nerve demonstrated by the CT scan in this case is also very unusual. These findings led us question the accuracy of the diagnosis of THS and prompted the performance of the biopsy. Classically, radiologic abnormalities in the THS have been restricted to carotid artery narrowing on angiography, occlusion of the superior orbital vein and sella turcica erosion on plain X-ray tomography. CT scan have generally been considered to be normal [1]. With improved resolution of CT scan and magnetic ressonance imaging studies, more recent studies have demonstrated the presence of enhancing lesions in the area of the cavernous sinus and superior orbital fissure, similar to the one we have observed in our patient [10,11]. Such findings could also have been caused by a number of other conditions including meningioma, carcinoma, metastasis, lymphoma, sarcoidosis and Wegener's granulomatosis [1,10,11]. Some of these conditions can present clinically as painful ophthalmoplegia and simulate the THS. Furthermore, the dramatic improvement with high dose corticosteroid therapy, that has been recommended as a diagnostic test for THS, has also been reported with other causes of painful ophthalmoplegia including neoplasms [1]. Therefore, it is important to emphasize that the THS should still be a diagnosis of exclusion.

RESUMO

Este trabalho relata um paciente de 65 anos, sexo masculino, com síndrome de Tolosa-Hunt que apresentou oftalmoplegia dolorosa unilateral e perda visual bilateral. A tomografia computadorizada mostrou uma lesão hipercaptante na fissura orbitária superior e no canal óptico que à biópsia revelou se tratar de processo inflamatório crônico inespecífico. Tratamento com corticosteróides resultou em melhora acentuada do quadro embora um olho tenha permanecido sem percepção luminosa. O processo inflamatório envolvia a fissura orbitária superior e o nervo óptico à direita e presumivelmente se extendia através da base do crânio para envolver o nervo óptico contralateral.

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