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The efficacy of radiation monotherapy for Tolosa-Hunt syndrome.

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Sirs,

Tolosa-Hunt syndrome (THS) is characterized by painful ophthalmoplegia due to an idiopathic chronic granulomatous inflammation of the cavernous sinus.[9] THS usually improves with corticosteroids, however, in cases showing an insufficient effect of corticosteroid therapy or side effects, alternative therapies such as immunosuppressive agents[3] or focal radiation therapy have been reported.[2,5-7] We describe a patient with THS who was successfully treated with radiation monotherapy.

A 36-year-old man with chronic hepatitis B and impaired glucose tolerance since age 30 developed acute left orbital pain and double vision at the end of July 2006. Neurologic examination on 3 August 2006 demonstrated only mild restriction of abduction of the left eye. Magnetic resonance imaging (MRI) of the brain showed no abnormality including the cavernous sinus region at that time. His illness progressed rapidly and he was admitted to our hospital on 14 August 2006.

On admission, body temperature and blood pressure were normal. Neurologic examination demonstrated almost complete ophthalmoplegia in all directions and

blepharoptosis without pupil abnormality on the left. Spontaneous left ophthalmic pain was also reported. There were no ophthalmic symptoms on the right and there were no symptoms of lower cranial nerves on both sides. Urinalysis and routine hematological examination were normal. Blood chemistry examination demonstrated mild elevation of aspartate aminotransferase (60 IU/L; normal range 11-34 IU/L), alanine aminotransferase (151 IU/L; normal range 6-39 IU/L) and glycohemoglobin A1c (6.1%; normal range 4.3-5.8 %). Surface antigen and core antibody of hepatitis B virus were positive. The following immunological and inflammatory investigations were normal or negative; antinuclear antibody, cytoplasmic and perinuclear antineutrophil cytoplasmic antibody, angiotensin converting enzyme level and β -D glucan. Chest radiography and Gallium scintigraphy of the whole body showed no abnormality suggestive of systemic inflammatory disease. Cerebrospinal fluid examinations were normal including cytology and culture for bacteria and mycobacteria. Brain MRI showed abnormal soft tissue enhanced with gadolinium within the left cavernous sinus (Figure).

Based on a diagnosis of THS, we first considered corticosteroid therapy for THS

with simultaneous antiviral therapy for hepatitis; however, the patient refused corticosteroid therapy because of the possible exacerbation of hepatitis. One week after admission, the left pupil was dilated and light response was diminished, indicating progression of THS. Thereafter, the patient received a total of 30 Gy in 15 fractions focused on the left cavernous sinus by three-dimensional conformal radiotherapy (3D-CRT).

Left blepharoptosis and ophthalmoplegia gradually improved within two weeks after the initiation of radiation therapy; blepharoptosis had almost completely disappeared by the middle of October 2006. Ophthalmoplegia also completely recovered by the beginning of January 2007. The illness remained in remission without any side effects during the observation period of one year.

However, the response to corticosteroid therapy could not be evaluated due to chronic hepatitis B, our patient fulfilled the remaining diagnostic criteria of THS by the International Headache Society,[4] and was successfully treated with radiation monotherapy.

Previous reports for radiation therapy in THS are shown in the Table.[2,5-7] All

four reported patients had received corticosteroid as an initial treatment, and radiation therapy was added because corticosteroid therapy showed insufficient effects (steroid dependence or steroid resistance) or severe side effects. It should be noted that our patient received only radiation therapy resulting in improvement of THS. To our knowledge, there has not been any previous report of radiation monotherapy for THS.

There has not been any consensus regarding the dose of radiation for THS. However, low dose radiation seems to be sufficiently effective as shown in the Table. The response of our patient demonstrated that low dose radiation monotherapy is effective in THS.

Additionally our patient and previously described patients who had received low dose radiation therapy for THS had no adverse effects related to the therapy during observation period (maximum 8 years). In case of exposure of the cavernous sinus to radiation, adverse effect on visual pathway and cranial nerves within the cavernous sinus should be concerned,[8] however, low dose irradiation to cavernous sinus by 3D-CRT method may be safe, referring to the neuronal tissue tolerance to irradiation (e.g. tolerance dose at the 5% complication rate within 5 years from treatment (TD 5/5)

for optic nerve and chiasma is 50 Gy).[1] Radiation monotherapy would be a useful and safe therapeutic option when it is difficult or contraindicated to use corticosteroids due to complications.

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References

1. Emami B, Lyman J, Brown A, Coia L, Goitein M, Munzenrider JE, Shank B, Solin LJ, Wesson M (1991) Tolerance of normal tissue to therapeutic irradiation. *Int J Radiat Oncol Biol Phys* 21:109-122.
2. Foubert-Samier A, Sibon I, Maire JP, Tison F (2005) Long-term cure of Tolosa-Hunt syndrome after low-dose focal radiotherapy. *Headache* 45:389-391.
3. Hannerz J (1992) Recurrent Tolosa-Hunt syndrome. *Cephalalgia* 12:45-51.
4. Headache Classification Subcommittee of the International Headache Society (2004) The International Classification of Headache Disorders; 2nd Edition. *Cephalalgia* 24(Suppl 1):1-160.
5. Larner JM, Malcom R, Miller JQ, Newman SA (1991) Tolosa-Hunt syndrome: A radiation responsive syndrome. *Neuro-ophthalmology* 11:289-292.
6. Mormont E, Laloux P, Vauthier J, Osseman M (2000) Radiotherapy in a case of Tolosa-Hunt syndrome. *Cephalalgia* 20:931-933.

7. Nasr YG, Boghen D, Vézina J (1987) Tolosa-Hunt syndrome with a prolonged course: Successful treatment with radiotherapy. *Neuro-ophthalmology* 7:57-60.
8. Tishler RB, Loeffler JS, Lunsford LD, Duma C, Alexander E 3rd, Kooy HM, Flickinger JC (1993) Tolerance of cranial nerves of the cavernous sinus to radiosurgery. *Int J Radiat Oncol Biol Phys* 27:215-221.
9. Tolosa E (1954) Periarteritic lesions of the carotid siphon with the clinical features of a carotid infraclinoidal aneurysm. *J Neurol Neurosurg Psychiatry* 17:300-302.

Figure legend

Axial sections of T1-weighted images before (A) and after enhancement with gadolinium (B). The soft tissue within the left cavernous sinus with intermediate signal intensity shows obvious gadolinium enhancement (arrow heads).

Table. Patients with Tolosa-Hunt syndrome treated with radiation therapy

Patient No.	Authors [reference]	Age (y.o)	Gender	Corticosteroid therapy	Immunosuppressant therapy	Dose of radiation (Gy)	Treatment period of radiation	Efficacy of radiation	Interval between initiation of radiation and improvement of the symptoms	Recurrence after radiation	Adverse effect related to radiation therapy	Observation period
1.	Nasr YG et al. (1987) [7]	49	female	yes	no	50	not available	yes	not available	no	not available	2 years
2.	Larner JM et al. (1991) [5]	50	male	yes	no	20	5.5 months	yes	not available	no	not available	11 months
3.	Mormont E et al. (2000) [6]	32	female	yes	yes (azathioprine)	30	22 days	yes	15 days	no	no	3 years
4.	Foubert-Samier A et al. (2005) [2]	41	male	yes	no	20	not available	yes	not available	no	no	8 years
5.	Present patient	36	male	no	no	30	21 days	yes	2 weeks	no	no	1 year

Figures

