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A Case of Tolosa-Hunt Syndrome Misdiagnosed as Migraine

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Abstract

A case report on Tolosa-Hunt syndrome misdiagnosed as migraine. A 21 year old woman presented with persistent unilateral headache and ocular pain. She visited a general hospital with headache and ocular pain, and underwent orbital and brain magnetic resonance imaging (MRI). However, there was no significant abnormality. She was diagnosed with migraine and relied on analgesics to control pain. After 30 days, ptosis and ophthalmoplegia began to develop. Her visual acuity was decreased in left eye, associated with 3rd, 4th and 6th cranial nerve palsy. Orbital MRI showed pathologic findings in the left superior rectus muscle and the orbit. With steroid therapy, extraocular muscle movement and visual acuity in left eye was improved. High signal in the superior rectus muscle and orbit decreased in follow-up orbital MRI. Tolosa-Hunt syndrome is a rare, benign condition, which should be excluded in patients with headache. MRI is essential to diagnosis and follow-up of this condition, although not confirmatory of this disease.

Key words : Extraocular muscles, Ophthalmoplegia, Tolosa-Hunt syndrome

INTRODUCTION

Tolosa-Hunt syndrome is a condition characterized by granuloma in the carvernous sinus or supraorbital fissure, associated with painful ophthalmoplegia, unilateral headache, sensory loss in the distribution of the ophthalmic division of the trigerminal nerve.[1] The diagnostic criteria for Tolosa-Hunt syndrome includes unilateral orbital pain, paresis, neurologic sign and responsiveness to steroid therapy. Contrast-enhanced MRI is the investigation of choice. The reported case herein showed atypical Tolosa-Hunt syndome patient who has more longer interval between clinical symptoms and onset of abnormal MRI findings rather than typical Toloca-Hunt syndrome As such, the authors combined the report with a literature review.

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CASE REPORT

A 21 year-old woman visited with persistent unilateral headache and ocular pain associated with nausea and dizziness, which began 40 days previously with unilateral headache and ocular pain. Since then, the symptoms had aggravated. 10 days after onset of headache, she had visited a local clinic. She underwent orbital and brain MRI which demonstrated no abnormality.(Fig. 1. A.) In addition, ophthalmic examinations were normal and she was diagnosed as migraine, for which she was sent to the pain clinic for follow-up and pain control.

Ptosis and ophthalmoplegia began to develop in the left eye 30 days following symptoms onset. Her initial corrected visual acuity (VA) was 1.25 / 0.4 (right / left), and the intraocular pressure was 17 / 15 mmHg. She complained of a painful diplopia and dizziness. On neurologic examinations, the left pupil was dilated to 4 mm compared to the right, at 3 mm. Her direct and indirect light reflexes were intact. Ocular movement examination demonstrated left-sided oculomotor, trochlear, abducent nerve palsy, and ptosis.(Fig. 2. A.) Fundus

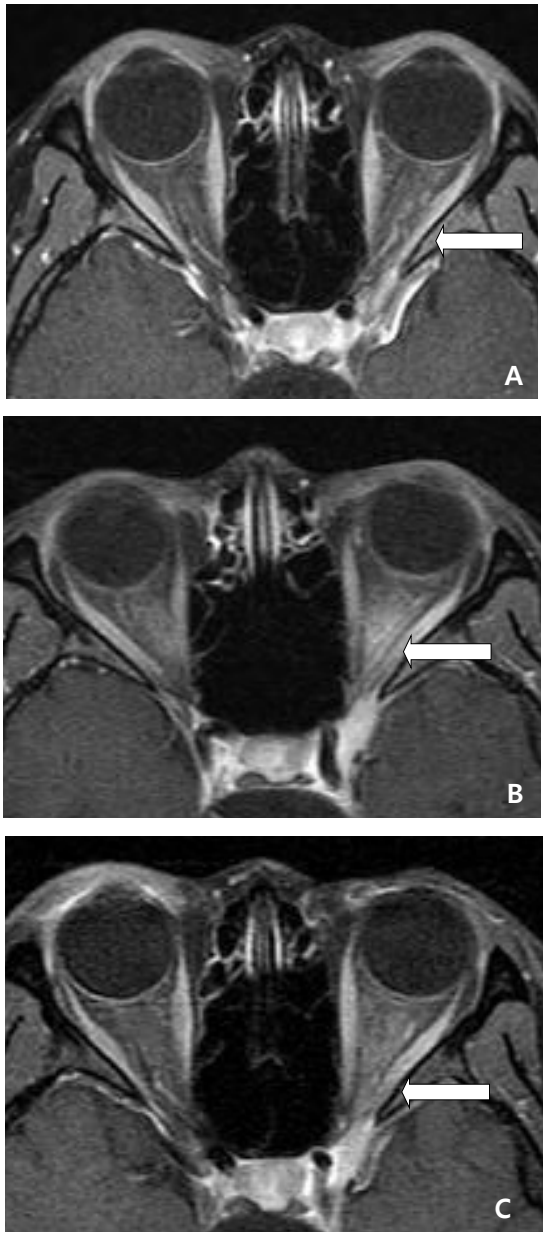
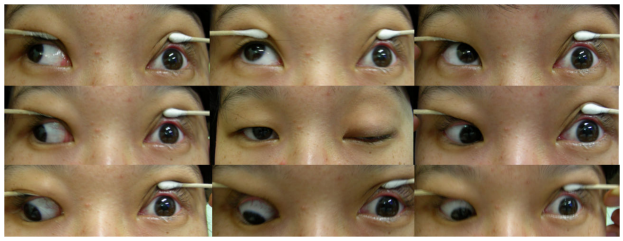
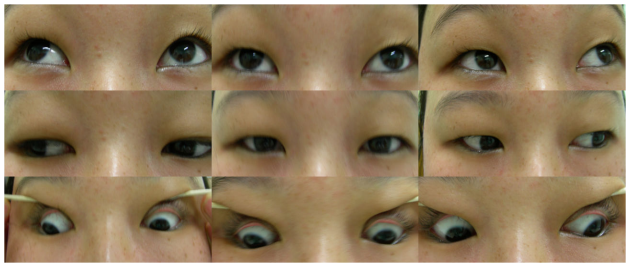


Fig. 1. Orbital MR images. (A): 10 days after the onset of ocular pain and headache, initial T1-weighted MR images shows normal superior rectus muscle or orbital structures. (arrow) (B): 40 days after onset of symptom, T1-weighted MR images of the left supeiro rectus muscle shows more enhanced findings than previous images. (arrow) (C) 1 months after stroid therapy, follow-up MR images shows reduced signals. (arrow)

examination was unremarkable and there was no pathologic reflex. Hematologic and biochemical profiles, including ESR, CRP and thyroid hormone were within the normal limits. HbsAg, VDRL, ANA and Anti ds-DNA Ab was negative. Orbital and brain CT showed normal finding but gadolinium enhanced orbital MRI showed high signal enhancement in the left superior rectus muscle and orbit without vascular abnormalities and space-occupying lesions. (Fig. 1. B.) VEP test showed delayed response in the left eye. CSF analysis was unremarkable.



(A)



(B)

Fig. 2. Cardinal movement. (A): 40 days after the onset of ocular pain and headache, Left ocular palsy and ptosis was developed. (B): With steroid therapy, the left ocular movement improved remarkably.

A diagnosis of Tolosa-Hunt syndrome was made in the light of the above findings. The patient was managed with systemic methylprednisolone 1000 mg for 6 days. 2 days after starting steroid therapy, her VA recovered to 1.0, and ocular pain as well as ptosis were significantly resolved. Subsequently, oral methylprednisolone 60 mg was given for 14 day, and gradually tapered at 10 mg weekly. Her extraocular muscle movement improved 2 weeks after initiating steroid therapy.(Fig. 3. A.) Follow-up VA assessment of the left eye was 1.25. Follow-up MRI demonstrated resolution of previous high signal enhancement in the superior rectus muscle and orbit.(Fig. 1. C.)

DISCUSSION

Several conditions can cause painful ophthalmoplegia. They including vascular, neoplastic, inflammation and infective causes. Treatment and prognoses of painful ophthalmoplegia are variable with the causes. It is therefore important to be able to establish the differential diagnoses of painful ophthalmoplegia.[2)]

Tolosa-Hunt syndrome was first described by Tolosa in 1954.[3)] It is a rare disorder and the exact mechanism is not unclear. It caused by non-specific granulomatous inflammation in the carvernous sinus, superior orbital fissure or orbital apex. Its characteristic symptoms include unilateral ocular pain and headache, sensory loss in the distribution of the ophthalmic and occasionally the maxillary division of the trigerminal nerve.[1)] Its incidence was 1 to 2 per million. And it may affect any age and no sexual predilection.[1)]

The International Headache Society (HIS) presented new diagnostic criteria for Tolosa-Hunt syndrome in 2004.[4)] The secriteria includes (A) one or more episodes of unilateral orbital pain persisting for weeks if untreated; (B) paresis of one or more of the third, fourth and/ sixth cranial nerves and/or demonstration of granuloma by MRI or biopsy; (C) paresis coincides with the onset of pain or follows it within 2 week; (D) pain and paresis resolve within 72 hours when treated adequately with corticosteroids; (E) other causes have been excluded by appropriate investigations.[4-5)]Basedonthenewclassification,our patient fulfills all the HIS criteria, except (C), because her paresis occurred later. We excluded viral and bacterial infection, cerebrovascular disease and tumor by serologic and neuro-imaging studies.[6)] Orbital MRI before and after steroid treatment helped diagnose the Tolosa-Hunt syndrome, which markedly respond to corticosteroid.[7)]

Usually ophthalmoplegia occurs within 2 weeks following onset of ocular pain or headache. However, in this case ophthalmoplegia began 4 weeks after ocular pain onset.[8)] In addition, there was no significant findings on early orbital MRI, which led to the initial misdiagnosis of migraine and delayed appropriate management. It may be an atypical case of Tolosa-Hunt syndrome - it is unclear why symptoms were delayed and early MIR findings were normal. Generally, contrast-enhanced MRI is accepted as an essential investigation modality. However, normal MRI findings do not completely exclude Tolosa-Hunt syndrome.[10-11)] As in this patient, careful follow-up and analysis of the patient's complains about persistent unilateral headache and ocular pain were vital.

CONCLUSION

Tolosa-Hunt syndrome is a condition characterized by granuloma in the carvernous sinus or supraorbital fissure, associated with painful ophthalmoplegia, unilateral headache, sensory loss in the distribution of the ophthalmic division of the trigerminal nerve. There is he diagnostic criteria for Tolosa-Hunt syndrome includes unilateral orbital pain, paresis, neurologic sign and responsiveness to steroid therapy. Generally, contrast-enhanced MRI is accepted as an essential investigation modality. But it may be an atypical case of Tolosa-Hunt syndrome and normal MRI findings do not completely exclude Tolosa-Hunt syndrome. So, careful follow-up and analysis of the patient's complaint about persistent unilateral headache and ocular pain were vital.

국문초록

단순 편두통으로 오진된 톨로사 헌트 증후군을 보고하는 바이다. 21세 여성이 지속적인 좌측 편두통 및 안구통을 주소로 내원하였다. 환자는 내원 40일 전 두통과 안구통으로 타 병원을 내원하여 안와 및 두부 MRI 시행

하였으나 특이소견이 발견되지 않았다. 증상 발생 30일 후 좌안의 안검하수와 안근마비가 발생하였다. 3번, 4번, 6번 뇌신경 마비와 함께 좌안의 시력저하가 동반되었다. 안와 MRI에서 좌안 상직근과 안와내에 이상소견이 발견되었다. 스테로이드 치료와 함께 외안근 기능과 시력이 호전되었고, 안와 MRI에서 상직근과 안와내에 보이던 고신호 강도도 감소하였다. 톨로사 헌트 증후군의 발병빈도는 낮지만, 만약 환자가 두통을 호소할 경우 반드시 배제해야하는 양성 경과를 보이는 질환이다. MRI는 톨로사 헌트 증후군의 확진 검사로서 한계점은 있으나, 진단 및 경과관찰을 위해 필수적이다.

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