

Contents lists available at ScienceDirect

Turkish Journal of Emergency Medicine

journal homepage: http://www.elsevier.com/locate/TJEM



Case Report

A case of headache, double vision and ptosis in emergency department: Tolosa-Hunt syndrome



Ataman Kose ^{a, *}, Seyran Bozkurt ^a, Sermin Tok Umay ^b, Serkan Karakulak ^a, Arda Yilmaz ^c

- ^a Mersin University, Faculty of Medicine, Department of Emergency Medicine, Mersin, Turkey
- ^b Mersin University, Faculty of Medicine, Department of Radiology, Mersin, Turkey
- ^c Mersin University, Faculty of Medicine, Department of Neurology, Mersin, Turkey

ARTICLE INFO

Article history:
Received 10 August 2017
Received in revised form
9 October 2017
Accepted 18 October 2017
Available online 31 October 2017

Keywords: Headache Double vision Ptosis Steroid treatment

ABSTRACT

Headache and double vision symptoms can be seen especially neurological and optical problems. We present Tolosa-Hunt syndrome in a 34-year-old female patient who presented to the emergency department (ED) with the complaints of unilateral severe headache, double vision and ptosis, presented asymmetric contrast enhancement in the right superior cavernous sinus on MR. This pain was reduced with steroid therapy. Pulse steroid therapy of 1 mg/kg/day was planned based on the patient's clinical manifestation, MRI findings and presumed diagnosis of THS. It was observed at patient's follow-up that there were occasional headache, ptosis and double vision were disappeared and MRI findings were regressed.

In the differential diagnosis of patients presenting to the ED with the complaints of headache and double vision, rare pathologies such as THS should be considered in addition of common pathologies such as neurological and ocular diseases.

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1. Introduction

Headache and double vision are common symptoms in ED. In the differential diagnosis of these symptoms, problems affecting different systems, especially neurological and optical problems can be seen. Among these, Tolosa-Hunt syndrome (THS), which is a rare disease, has been reported only as case series. THS is a painful ophthalmoplegia pathology which is often localized in cavernous sinus and manifested with involvement of the third, fourth, and sixth cranial nerves accompanied with unilateral headache. Herein, we present THS case in a 34-year-old female patient who presented to the ED with the complaints of unilateral severe headache, double vision and ptosis, presented asymmetric contrast enhancement in the right superior cavernous sinus segment on MRI and whose pain was reduced with steroids therapy.

E-mail address: ataberk76@yahoo.com.tr (A. Kose).

Peer review under responsibility of The Emergency Medicine Association of Turkev.

2. Case

A 34-year-old female patient presented to the ED with the complaints of headache and double vision. In her medical history, the patient described a painful right side migraine which occasionally occurred for five days and exacerbated on the day of her admission to the ED. There was also double vision with pain. In addition to these complaints, the patient noticed ptosis on the day of admission. There was no any disease in her history except for subclinical hyperthyroidism. On her examination, general status was good, conscious was open and the patient was oriented and cooperated. Vital signs were found as blood pressure: 110/80 mm/ hg, pulse: 66/min, temperature: 36.8 °C and oxygen saturation: 99%. On her neurological examination, bilateral light reflex was pupillary isochoric. There were ptosis in the right eye, double vision in the right eye looking upward and leftward and 3rd cranial nerve paresis. Hypoesthesia was identified in dermatomas of the ophthalmic and maxillary branches of the trigeminal nerve in the right half of her face and corneal reflex could not be detected on the right. No lateralize muscular deficits was noted and the sitting balance was evaluated as normal. The other system examination of the patient was natural. After establishment of the vascular access, hydration, metoklopramid HCI 10 mg/2 ml and analgesics were

^{*} Corresponding author.

given to the patients and blood tests were ordered. No abnormal findings were found in the blood tests of the patient. In addition, the patient's lipid profile, thyroid function tests, sedimentation and anemia profiles (iron, ferritin, folic acid, and vitamin B12) were in normal ranges. The patient underwent cranial computed tomography (CT) in the ED. The patient with CT evaluated as normal was consulted and admitted to the neurology clinic. On the cerebral MRI of the patient, a focal soft tissue lesion showing asymmetrical contrast enhancement to the posterior tentorial surface and having not clear margins was observed in the right superior cavernous sinus section (Fig. 1). THS Pulse steroid therapy (Metilprednizolon sodyum suksinat, Prdenol-L) of 1 mg/kg/day was planned based on the patient's clinical manifestation. Steroid therapy starting with 1 mg/kg was recommended in the patient who benefited from the treatment so as to be reduced 0.5 mg each week. She was advised for neurology outpatient clinic control visit after one month and discharged. It was observed at patients' follow-up that there were occasional headache, ptosis and double vision were disappeared and MRI findings were regressed. MRI and positive response to the treatment with corticosteroids were relevant for making the diagnosis.

3. Discussion

Neuro-ophthalmologic emergencies are not fully described, but optic and visual symptoms are thought to be predominant as neurological emergencies that should be urgently evaluated and treated. In neuro-ophthalmologic emergencies, the emergency physician is confronted consultation with the relevant department, performing imaging studies and determining the need for hospitalization. Especially central and ophthalmologic causes, all



Fig. 1. Asymmetric enhancement (arrow) extending from the right cavernous sinus supraoposterior toward the tentorium on contrast enhanced axial MRI examination (arrow).

systems should be examined and investigated in patients presenting to ED with the complaints of headache and double vision. The International Headache Society published a report on other causes (tumors, vasculitis, basal meningitis, sarcoidosis, diabetes and ophthalmoplegic migraine) of painful ophthalmoplegia that should be considered in differential diagnosis of diagnostic criteria for THS. In our case also THS was considered after excluding the other causes through clinical, laboratory and imaging tools. The majority of cases reported as THS consisted of adults without sex discrimination. In addition, pain is usually unilateral, and double vision is mostly due to the influence on the 3rd cranial nerve. 4,6-8 Günaydın et al.⁹ and Hatipoğlu et al.⁴ reported unilateral headache, double vision and the involvement of the third and fifth cranial nerves. Similarly, in our study the pain was on the right side and the 3rd and 5th nerves were affected. In some studies, ptosis accompanies as in our study. 4,6–9 The diagnosis is usually made with contrast-enhanced MRI, as well as clinical and physical examination. It affects frequently the cavernous sinus as well as the orbital apex, and the superior orbital fissure. Significant contrast enhancement is seen after enlargement and contrast administration in the cavernous sinus in MRI. 1,5,9-11 Routine blood tests and cranial CT of our patients were normal. Asymmetric contrast enhancement was observed in the superior portion of the right cavernous sinus on cerebral MRI ordered. Though there is no clear consensus about treatment in THS patients, steroid therapy remains the first treatment. Ophthalmologic and neurological symptoms and signs, especially headache can be resolved with steroids therapy. In general, starting with 1 mg/kg pulse steroid is recommended as the treatment. Among the THS diagnostic criteria published by the Classification Committee of the International Headache Society in 2004, there was improvement of the clinical picture with steroid treatment. However, this criterion was omitted in the definition published in 2013. However, in most studies this definition is an important parameter.^{3,5,6,12} In a series of two cases reported by Günaydın et al.⁹ and Hatipoğlu et al.,⁴ symptoms and signs of the patients were improved with methylprednisolone therapy. In retrospective screening of 16 pediatric patients with THS, 15 were treated with steroids and the majority of complaints were reported to resolve within 72 hours.

In the present case study; diagnosis of THS was considered because of unilateral headache, double vision, ptosis and involvement of the 3rd and 5th cranial nerves, excluding of the majority of other causes, detection of contrast enhancement in the cavernous sinus on contrast enhanged MRI study and regressson of headache ptosis and double vision with steroid therapy. In the differential diagnosis of patients presenting to the ED with the complaints of headache and double vision, rare pathologies such as THS should be considered in addition of common pathologies such as neurological and ocular diseases.

Funding

None Declared.

Conflicts of interest

None Declared.

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