

TRAUMATIC OPTIC NEUROPATHY – CASE REPORT WITH DISCUSSION ON DIAGNOSTIC PROCEDURES AND THERAPY

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SUMMARY – Traumatic optic neuropathy (TON) is a serious vision threatening condition that can be caused by ocular or head trauma. Indirect damage to the optic nerve is the most common form of TON occurring in 0.5% to 5% of all closed head trauma cases. Although the degree of visual loss after indirect TON may vary, approximately 50% of all patients are left with 'light perception' or 'no light perception' vision, making TON a significant cause of permanent vision loss. We present a 47-year-old male patient with a history of right eye keratoconus following a motorcycle crash. Visual acuity was of 'counting fingers at 2 meters' on the right eye due to keratoconus and 'counting fingers at 1 meter' on the left eye as a consequence of trauma. The Octopus visual field showed diffuse reduction in retinal sensitivity and the Ishihara color test indicated dysfunction of color perception on the left eye. Relative afferent pupillary defect was also present. Computed tomography revealed multifragmentary fracture of the frontal sinus and the roof of the left orbit without bone displacement. Based on the findings, conservative corticosteroid therapy without surgery was conducted. The patient responded well to treatment with complete ophthalmologic recovery.

Key words: *Head injuries, closed; Motorcycles; Optic nerve injuries; Eye injuries; Visual fields; Visual acuity; Diagnostic imaging; Keratoconus; Treatment outcome; Case reports*

Introduction

Eye injuries and their consequences represent a significant public health problem. Trauma of the eye and its surrounding structures is one of the main causes of severe visual impairment and blindness, particularly in the younger male population¹⁻⁴. Although ocular trauma has generally been overlooked, it has been identified as a relevant cause of visual impairment with

blindness in an estimated 1.6 million cases as a result of eye injuries with additional 19 million having monocular blindness or low vision due to eye trauma¹.

Facial fractures are commonly associated with eye injuries resulting in different levels of vision loss. According to various studies, the estimated incidence of ocular damage after midfacial fractures ranges from 2.7% to 67%. As ocular dysfunction may cause permanent visual disability, any curable ocular trauma needs to be detected and treated as early as possible. Thus, all patients with orbital blow-out fracture associated with visual disability require prompt and efficient ophthalmologic consultation. In fact, posterior orbital fracture appears to be associated with worse visual repercussions than those located anteriorly⁴⁻⁸ (Table 1).

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Received September 2, 2016, accepted October 20, 2016

Table 1. Symptoms and signs requiring immediate ophthalmologic intervention in patients with head or facial trauma

Function and findings	Symptoms and signs
Visual acuity	Reduced/loss of vision
Pupillary size	Dilated
Pupillary reactions	Sluggish or loss of direct reflex Sluggish or loss of consensual reflex
RAPD (swinging light test)	Present
Pain	Present
Color test	Reduced color perception
Color of the eye	Loss of red reflex
Position of the globe	Proptosis Enophthalmus
Eye movement	Reduced Diplopia Accompanied by pain
Anterior orbital fractures	Poorer visual outcome

RAPD = relative afferent pupillary defect

Traumatic optic neuropathy (TON) is a serious vision threatening condition that can be caused by either ocular or head trauma and is conventionally classified into direct and indirect injury⁸. Direct TON is caused by injury precisely to the optic nerve area causing dysfunction and anatomic disruption. It is usually associated with severe visual loss with a minimal chance for recovery. Indirect TON is caused by acceleration and deceleration forces due to blunt head or closed globe trauma. This type of TON preserves ocular and cerebral tissue, however, it indirectly disrupts the functional and anatomic integrity of the optic nerve with vision loss varying from mild to total blindness⁸⁻¹⁰. Indirect damage to the optic nerve is the most common form of TON occurring in 0.5% to 5% of all closed head trauma cases¹⁰, as well as in 2.5% of those with midfacial fractures¹¹. Clinical diagnosis of indirect TON is based on evidence of optic nerve dysfunction in patients having sustained craniofacial trauma, with normal appearance of the optic nerve head on clinical retinal examination^{8,9}. The most common site of indirect TON is the part of the optic nerve situated in the optic canal, followed by the intracranial part of the optic nerve and chiasm². Although the degree of visual

loss after indirect TON may vary, approximately 50% of all patients are left with 'light perception' or 'no light perception' vision, making TON a significant cause of permanent vision loss^{8-10,12}. The pathogenesis is still ambiguous with several possible mechanisms responsible for actual visual disability. The mechanism of TON occurrence can be divided into primary and secondary. Primary mechanism is mechanical shearing of the optic nerve axons and contusion necrosis due to immediate ischemia from damage to the microcirculation. Secondary mechanism occurs *via* apoptosis of both injured and initially unharmed adjacent neurons^{8,9,12}.

Indirect TON may be treated with various doses of steroids, or alternatively surgical optic canal decompression can be performed^{2,4,8,10}. However, at present, there is no proven mode of treatment for this condition, with continuing controversy over optimal standard modality. The International Optic Nerve Trauma Study (IONTS) group found no significant difference in visual acuity outcome between treated and untreated patients, with the conclusion that neither form of available treatment can be recommended as the preferred option. Therefore a clinically reasonable decision on whether or not to treat and which mode of treatment to administer should be viewed on an individual patient basis¹³. Thus, in the absence of controlled guidelines, correct management of TON still remains a clinical dilemma.

Case Report

A 47-year-old male patient with a history of right eye keratoconus two hours following a motorcycle crash was admitted to the hospital. He displayed no signs of any brain injury and remained conscious without dizziness or vomiting. Since childhood, he had keratoconus of the right eye, which he never attempted to correct.

External examination showed pronounced upper and lower lid hematoma with large linear laceration in the left side of the forehead above the eyebrow. Ocular examination revealed visual acuity of 'counting fingers at 2 meters' on the right eye due to keratoconus and 'counting fingers at 1 meter' on the left eye on Snellen chart. Subconjunctival hemorrhage of the nasal conjunctiva in the left eye and relative afferent pupillary defect (RAPD) were observed. The Octopus visual field showed pronounced diffuse reduction in retinal

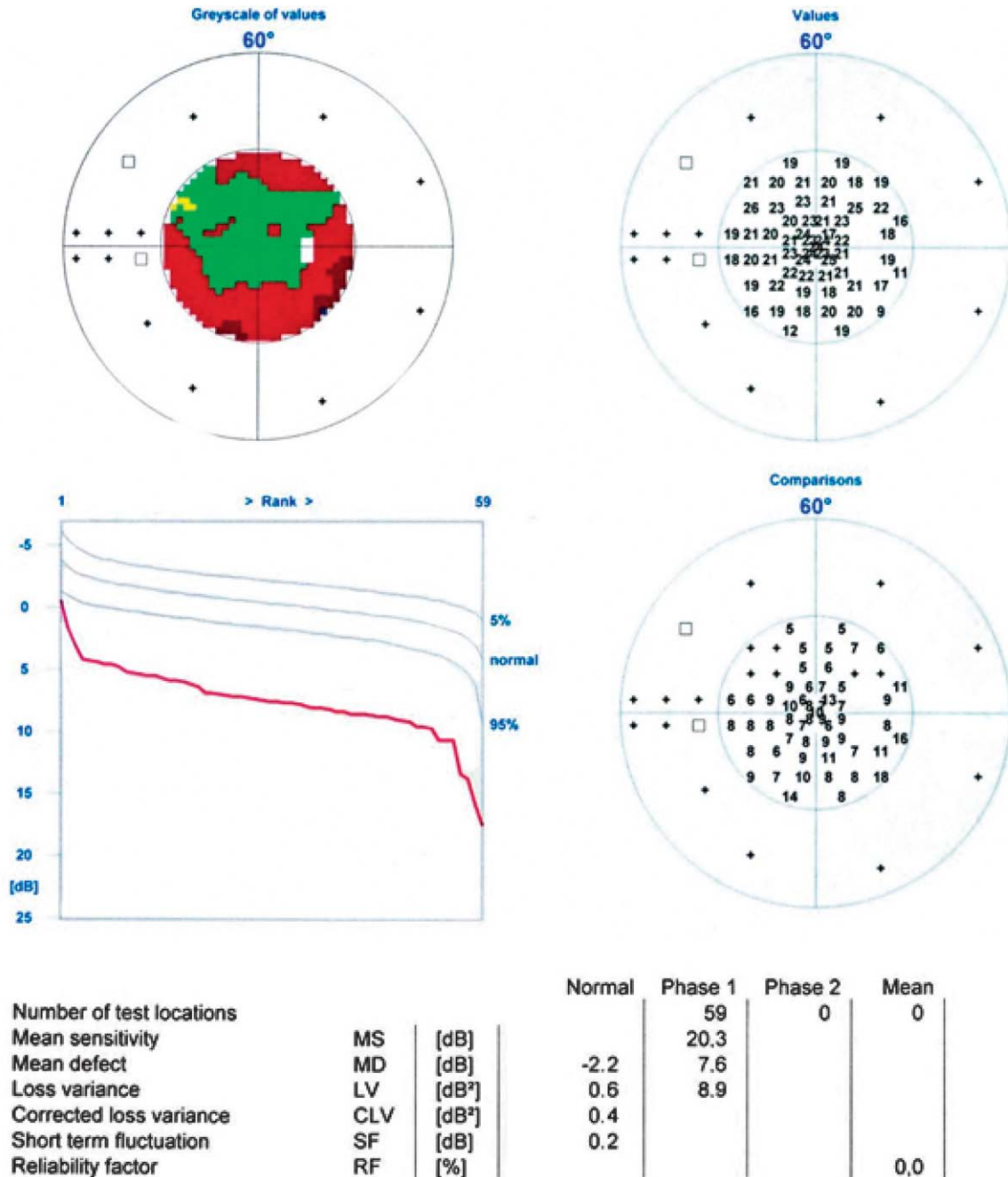


Fig. 1. Octopus visual field of the left eye.

sensitivity on the left eye (Fig. 1), whilst the finding for the right eye was within the normal limits (Fig. 2). Fluorescein staining of the cornea was negative, the anterior chamber and ocular lens were clear, and the intraocular pressure was normal. The retina and optic nerve head appeared normal on funduscopy with extraocular movements of both eyes within the normal

limits. Ishihara subjective color test indicated dysfunction of color perception of the left eye. Non-contrast computed tomography (CT) revealed a multifragmentary fracture of the frontal sinus and the roof of the left orbit without bone displacement.

Based on the findings, conservative medical therapy without surgery was conducted. The patient under-

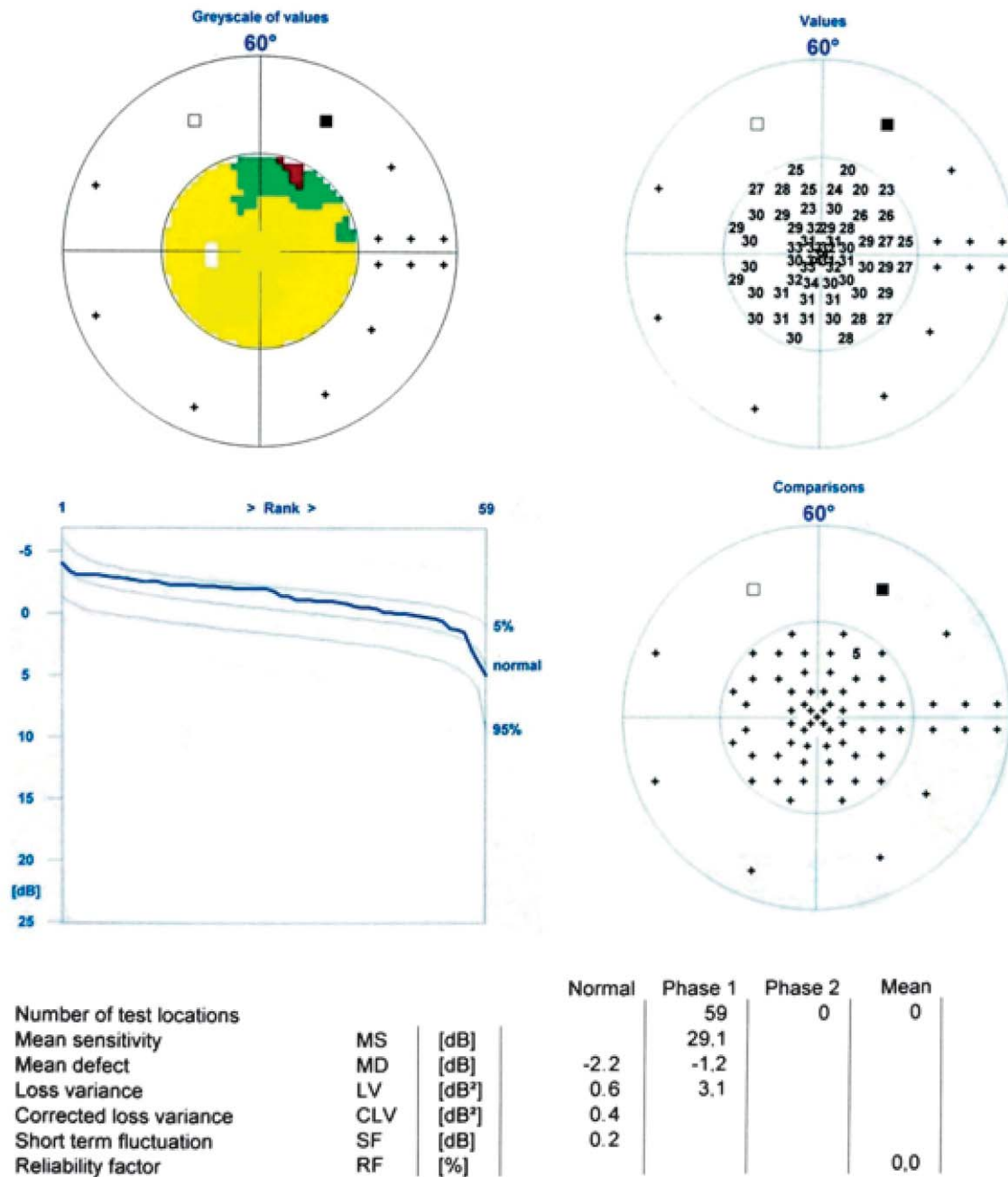


Fig. 2. Octopus visual field of the right eye.

went steroid treatment with intravenous application of methylprednisolone 250 mg *q.i.d.* for 3 days, followed by oral prednisolone 1 mg/kg for the next 11 days¹³. On the second day of therapy administration, visual acuity began to improve with the best visual acuity (BCVA) being 0.2, and then 0.5 and 1.0 on the 4th and 6th day, respectively. Visual field and Ishihara subjective

color tests also showed improvement during the period of hospital stay. The patient was discharged with normal ophthalmologic status and the recommendation for correction of keratoconus of his right eye. Ophthalmologic findings, including pupil reactions, fundus examination, optic disc and Octopus visual field were within the normal physiological limits on

each check up during two-year follow up. However, he did not make any effort to correct keratoconus of his right eye throughout this period.

Discussion

Blunt ocular trauma may cause various effects and damage all segments of the eye. It may be isolated or more frequently occur as part of head trauma, particularly facial area encompassing the zygomatic bone and maxillary sinus. The reported incidence of ocular injuries in patients with orbital fractures varies widely from 2.7% to 90%. Optic neuropathy is potentially a blinding complication that accompanies head or orbital trauma. It represents a frequent and preventable cause of visual impairment^{7-9,12}, with prompt diagnosis and management being essential in order to prevent vision loss. The most common form of TON is indirect damage to the optic nerve with the reported incidence varying from 0.5% to 5% of all closed head trauma cases¹¹. Damage to the optic nerve can be intraorbital, intracanalicular or intracranial, with the possible causes being hematoma, ischemia or direct bone fragment penetration. In addition, other traumatic retinal damage including edema, as well as ciliary artery impairment may lead to ischemia of the optic nerve with all its consequences. The clinical presentation of TON may vary widely, whereby the degree of visual impairment is not always proportional to the severity of trauma. Approximately 50% of patients are left with 'light perception' or 'no light perception' vision, suggesting TON as a significant cause of permanent visual loss. It has been shown that the mechanism of injury is a stronger predictor of final visual outcome than the fracture pattern itself^{4,15}. In a retrospective study of 35 patients, Carta *et al.* correlated poor outcome in patients with TON with the presence of blood in the posterior ethmoid cells, loss of consciousness, age over 40, and absence of improvement after two days of steroid treatment¹⁰.

Traumatic optic neuropathy is an uncommon yet potentially serious complication since optic nerve contusion or compression can result in total vision loss in healthy looking eyes. Visual acuity in patients with indirect TON may be significantly reduced^{8,9,15,16}; however, most of these ocular injuries are transient with no permanent consequence. Nevertheless, TON represents one of the real ophthalmologic emergencies. The

initial goal in treating TON is early recognition since the window of opportunity for efficient treatment may be less than 8 hours¹². Treatment choice with immediate surgical decompression of the optic nerve or the application of high doses of corticosteroids still remains debatable^{8,9,12,13,15,16}. Corticosteroids were used initially to decrease edema and vasospasm in an effort to limit ischemic nerve cell death. The rationale for intravenous corticosteroids in the treatment of TON was derived from the results of the NASCIS II^{11,17,18}. The NASCIS I, II and III showed benefits of therapy in patients with spinal cord injuries who received high-dose corticosteroids within 8 hours^{7,14}. Although they are a widely accepted form of therapy, their advantage in TON treatment has not yet been proven. In fact, the benefit of any kind of intervention is yet to be established⁷. Several studies failed to show clear benefit of corticosteroid therapy or optic nerve decompression, concluding that neither of the above mentioned therapies should be considered as standard care for patients with TON. Therapeutic decision should be based on an individual approach, bearing in mind the possible harmful side effects, as well as clinical benefit of the chosen treatment¹³.

In diagnostic procedures, CT is the neuroimaging study of choice for visualizing the bones of the optic canal, the paranasal and frontal sinuses. It is used to eliminate intraocular or orbital foreign bodies, as well as to detect acute orbital or intracranial hemorrhages. Alternatively, magnetic resonance imaging is the preferred method for visualizing soft tissue and is superior to other radiological examinations due to its three-dimensionality. Furthermore, it enables better evaluation of the orbital apex, determination of the presence of cavernous sinus damage, as well as the presence of hematoma of the optic nerve and detection of non-metallic foreign bodies¹⁹.

Since it is well known that TON may be the cause of severe visual impairment, we present the case of a patient with keratoconus of the right eye who is practically monocular due to his own negligence. He never made an attempt to correct his visual impairment regardless of the fact that he has been aware of his condition since childhood. The experience of trauma on his left healthy eye and consequential severe visual impairment left him practically blind for some period. It was one of the reasons for using corticosteroid therapy despite the current controversies in the treatment of

TON. Considering that the patient arrived to the hospital within two hours of injury, the possibility of early therapy and his age over 40 were additional reasons for the choice of treatment. Furthermore, he was a relatively healthy individual. In this case, the selected treatment was successful with complete recovery of visual acuity on the patient's left eye.

In conclusion, appropriate options for severe TON treatment are still controversial, however, the most widely accepted protocol includes the following: diagnosis of TON based on reduced visual acuity and presence of RAPD, administration of high doses of intravenous corticosteroids in the early stage of the disease, and switching to oral administration after 48–72 hours of treatment with constant monitoring of visual acuity and RAPD. Alternatively, in the absence of improvement in clinical parameters, operative decompression of the optic nerve is recommended^{8,9,14,20}. It should be emphasized that all patients sustaining head trauma, particularly facial and orbital trauma require complete ophthalmologic examination and evaluation with visual acuity and RAPD being monitored during all phases of treatment and recovery.

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Sažetak

TRAUMATSKA OPTIČKA NEUROPATIJA – PRIKAZ SLUČAJA
S RASPRAVOM O DIJAGNOSTIČKIM POSTUPCIMA I LIJEČENJU

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Traumatska optička neuropatija (TON) može biti uzrokovana traumom oka ili glave i predstavlja ozbiljno stanje koje može ugrožavati vidnu funkciju. Indirektno oštećenje očnoga živca je najčešći oblik TON-a, a javlja se u 0,5% do 5% slučajeva svih zatvorenih trauma glave. Iako stupanj gubitka vida nakon indirektno traumatske vidnoga živca može varirati, u oko 50% svih bolesnika vidna oštrina je smanjena na “osjećaj svjetla” ili “bez osjećaja svjetla”, zbog čega TON predstavlja značajan uzrok trajnog gubitka vida. Prikazan je slučaj 47-godišnjeg bolesnika s povredom vidnoga živca nakon motociklističke prometne nezgode. Vidna oštrina na desnom oku bila je “brojanje prstiju na 2 metra”, zbog otprije postojećeg keratokonusa i “brojanje prstiju na 1 metar” na lijevom oku kao posljedica traume. Nalaz vidnoga polja Octopus pokazao je difuzno smanjenje osjetljivosti mrežnice, a test Ishihara za boje ukazao je na poremećaj osjeta boja na lijevom oku uz prisutnost relativnog aferentnog pupilarnog defekta. Kompjutorizirana tomografija pokazala je multifragmentne frakture frontalnog sinusa i krova lijeve orbite bez pomaka kostiju. Na temelju kliničke slike i nalaza provedeno je uspješno konzervativno liječenje kortikosteroidima s potpunim oftalmološkim oporavkom.

Ključne riječi: *Glava, ozljede, zatvorene; Motocikli; Optički živac, ozljede; Očne ozljede; Vidna polja; Vidna oštrina; Dijagnostičko snimanje; Keratokonus; Ishod liječenja; Prikazi slučaja*