Traumatic Optic Neuropathy

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Abstract

Traumatic optic neuropathy is a rare pathology caused by an acute injury of the optic nerve secondary to trauma. The diagnosis is mainly clinically based on history and ophthalmic signs. Images sometimes show a direct lesion but most frequently the mechanism is indirect and affects the intracanalicular portion of the nerve. Incidence is higher in young males. Different treatments have been proposed including corticosteroids and surgery but the data in the literature to date has not shown any treatment to be superior to observation. Moreover, 40–60% of TON have spontaneous recovery, excluding severe cases.

Keywords

Trauma · Direct, indirect mechanism
Multifactorial · Spontaneous recovery

17.1 Introduction

Traumatic optic neuropathy (TON) refers to an acute injury of the optic nerve secondary to trauma. The diagnosis of TON is made clinically, based on history and ophthalmic signs. Like other optic neuropathies, patients with TON may have decreased central visual acuity (VA), decreased color vision, an afferent pupillary defect, and/or visual field (VF) defects.

17.2 Definition

TON is an optic nerve (ON) injury as a result of trauma, which results in total or partial loss of function, which may be transient or permanent. Trauma can lead to direct or indirect aggression to the nerve, the latter being the most common.

It may be due **to severe, moderate, or even mild head trauma**. The gravity varies according to different series; **43 to 56% have a severe visual loss,** but we must keep in mind that mild and/or spontaneously improved cases are often not reported. A vast majority are unilateral, but it can be bilateral if it involved major trauma. Since it involves head trauma, management should be carried out by a multidisciplinary team that includes emergency room doctors, head and neck surgeons, neurosurgeons, and ophthalmologists. Ophthalmological assessment should be carried out at the first possible opportunity, after stabilization of vital functions [1, 2].

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17.3 Incidence: Etiology

Is a rare pathology; it occurs in 1 to 5% of closed traumas. Its highest incidence is in young males: in *the International optic nerve trauma study,* the average age of involvement was 34 ± 18 years and 85% were male.

The most common cause is accident with motor vehicles or bicycles (49%), falls (27%), and aggression (13%). In pediatric series, the most common cause are falls (50%) followed by traffic accidents (40%). The most common clinical form is indirect form.

17.4 Classification

In accordance with the mechanism of action:

- **Direct:** There is a significant anatomical alteration of the optic nerve, for example, by a projectile that penetrates orbit at high speed, or as a result of avulsion of the optic nerve, which usually occurs with severe visual loss.
- **Indirect:** Forces that occur during the blow are transmitted from a distant site to the ON, which occurs in 0.5 to 2.5% of all closed head trauma, without any manifest damage to the structures of the surrounding tissues. The deforming tension transmitted to the skull of closed trauma is concentrated in the optic channel region.

According to the site of the injury:

- Head of the ON
- Intraorbital
- Intracanalicular
- Intracranial

The **intracanalicular portion is** the most likely to be injured in closed trauma, as the upper half of the nerve in the canal is strongly attached to the periosteum by the fusion of the nerve with the nerve sheath, and therefore has very reduced mobility making it especially susceptible to trauma. Direct nerve damage or ischemic damage due to vascular injuries may occur. Imaging studies, particularly computed tomography, show variable results—up to 50% according to some series—in relation to detecting fractures in the optical channel that are difficult to highlight. Sometimes it is possible to detect small bone fragments that can completely or partially section the nerve, or compress it.

The **intracranial part of** the ON begins at the optic channel output and reaches the chiasma. The sickle dural fold is very close and also fused to the nerve sheath, so injury may occur at this level. It is the second most common location of TON. The chiasmatic region is the third in frequency of involvement.

In accordance with the commitment of Fundus Oculi - FO-:

- **Previous** neuropathy: Lesions prior to the entry of the central artery of the retina into the ON, about 1 cm from the back of the eyeball, cause alterations in the FO: hemorrhage or edema, due to arterial or venous compromise. Optical nerve avulsion may also be observed in severe trauma.
- **Subsequent** neuropathy: FO is normal which makes diagnosis difficult. If the trauma causes definitive damage, partial, or total paleness will become apparent late, within a variable period of 3 to 6 weeks.

17.5 Pathophysiology

In direct **TON** the mechanism is evident and neuroimaging usually shows direct damage to the optic nerve or compression by a bone fragment or hematoma [3].

In indirect **TON** the mechanism is usually **multifactorial** with a combination of mechanical and vascular events. The biomechanical response of cranial content to trauma is an important component of the mechanisms that are generated. Holographic studies with laser interferometry of the orbital cavity have shown that biomechanical forces triggered by trauma are transmitted through the wall of the orbital cavity and produce maximum deformity at the vertex level. Elastic deformation of sphenoids is manifested by the

appearance of canal fractures. The complex arrangement of collagen fibers in the ON pods and their firm bonding to the canal allow stress forces to be transmitted to the nerve, with a longitudinal orientation. When intense, they cause stretching of nerve fibers. In addition, there is compression and rupture of the nutrient vessels in their journey through the scissor pattern of the collagen fibers of the dura mater, when shearing forces are generated, resulting in the appearance of microinfarctions and microhemorrhages with edema; this, in turn, further compromises the blood supply in an inextensible space such as the bone walls of the canal [4].

Primary damage **occurs to** axons in ganglion cells, which are irreversibly injured with consequent cell degeneration. Secondary mechanisms of edema, **especially significant ischemia** within the narrow optic channel are also triggered. This creates a compartment syndrome and a vicious circle between the two mechanisms involving multiple metabolic pathways—release of free radicals, alteration of calcium metabolism, alteration of vasoregulatory mechanisms—leading to death by apoptosis of affected ganglion cells. Postulated treatments aim to try to curb this secondary damage.

Diffuse **axonal damage should also** be considered as an underlying mechanism in indirect TON. Axon deformations occur in brain white tissue tracts that damage the axonal cytoskeleton resulting in impaired axoplasmic transport.

On the other hand, ophthalmologic controls should be maintained because visual decrease may not occur immediately to trauma but over time, due to the **development of intraorbital hematomas** or in the sheath of the optic nerve. Orbital hemorrhage results in compartment syndrome with increased orbital pressure that compromises the circulation of the ON. Orbital **emphysema,** after fractures, is usually a benign condition. However, cases have been reported in which a valve mechanism is generated, with a large increase of intraorbital pressure and ON compression, although it is an extremely rare form of TON.

The least common variant of TON is **ON avulsion.** It usually occurs after closed-balloon trauma and can be total or partial. Three likely production mechanisms have been proposed:

- Sharp increase in intraocular pressure from traumatic compression that disinserts the ON. It behaves similarly to an eye rupture at the level of an anatomically weak site in the eye wall.
- Increased orbital pressure with balloon protrusion, resulting in a stretching of the nerve, until it is avulsed from its scleral insertions.
- Extreme rotations of the globe with disruption of the laminar region.

17.6 History

Clinical **history is, of course, fundamental.** The type of trauma, the time of evolution, whether or not there was loss of consciousness, and the duration of the trauma should be recorded. If there was a visual loss, if it was perceived immediately or deferred. If the patient has received any treatment or surgery.

In mild cases, the patient may have difficulty referring trauma as he/she does not link it to late visual damage. In severe cases, particularly loss of consciousness, it is difficult to obtain both a history and the patient's examination.

A complete **ophthalmological examination should always be performed for signs and symptoms that** may accompany TON: edema—bruising—exophthalmos—obvious wounds or fractures—commitment to motility—and, except contraindication, FO with dilation to rule out, for example, retinal detachment, vitreous hemorrhage, intraocular foreign body, choroid ruptures, or crystalline dislocation. A history consistent with TON would be vision loss after blunt or penetrating trauma that could not be explained by slit lamp or dilated fundus findings.

In **physical examination,** it is essential to feel the orbital rim to look for signs of fractures. When orbital compartment syndrome is suspected, the existence of resistance to eye backpulsion and intraocular pressure should be explored which, if elevated, point to such diagno-

sis. The ocular globe and its annexes should be examined for concomitant eye disturbances.

1. **Decreased Visual Acuity:** The compromise is variable and ranges from non-light perception to minimum or imperceptible loss. Up to 40–60% of patients have a severe loss. Direct TON with very small VA has a worse prognosis. In the case of indirect injuries, recovery may be greater, and the initial VA has proven to be the most important prognostic factor: the higher the initial VA the greater the chance of improvement.
2. **Afferent pupillary defect**: Pupil evaluation is critical and should always be recorded in clinic history, particularly in cases with normal FO where the patient refers to marked visual disorder. On the other hand, in a patient with severe head trauma the onset of uni- or bilateral mydriasis implies the possibility of brain herniation with life risk. In bilateral cases, we can find diminished or abolished pupillary reflexes.
3. **Alteration in chromatic vision**: The blue–yellow axis can be affected with good VA and respected VF **or** red–green axis, when there is great visual affectation and damage of the central VF. Diffuse defects without a defined axis may also occur, and when the VA is severely affected, it is not possible to evaluate color vision correctly [5].
4. **Variable campimetric defects**: Lower hemianopsia (by involvement of upper pial vessels in the optic canal) and central dense scotomas are common.
5. **Fundus oculi:** The compromise is variable depending on the location of the insult. It is noteworthy again that a normal FO does not rule out TON as paleness takes time to become apparent.

17.7 Complementary Examinations

- **Visual field.**
- **OCT**: Allows follow-up injuries. When there is FO damage, it will show the thickness increasing of the RFNL and/or macula by edema and bleeding and its subsequent evolution toward normalization or atrophy.
- In indirect TON, defects will not become evident for 3 or 4 weeks, when a reduction in thickness in the peripapilar RFNL can be seen. The loss will stabilize at 20 weeks [6].
- **Orbital ultrasound** has limited utility restricted to anterior lesions.
- **Visual Evoked Potentials (VEPs)** are generally of limited utility because of the impossibility of mobilization in most cases. In patients with amaurosis and normal FO, highly altered or non-recordable VEPs allow to establish the extent of the damage; the possibility of visual recovery is highly unlikely.
- **NEUROIMAGES**: They are fundamental.
- **Computed Tomography** (**CT**) allows better detection of cranial fractures, and in particular, those that affect the optic channel. It is the most accessible and easiest study to perform even in severe traumas with loss of consciousness. It also allows to discard metallic foreign bodies, emphysema, bone fragments and, although with less definition, hematomas.
- **Magnetic Resonance Imaging (MRI)** allows for better assessment of brain matter, (presence of edema or stroke), as well as its prognosis when performing diffusion and perfusion techniques; it also detects signs of intracranial hypertension syndrome in particular cerebrospinal fluid in the optic nerve sheath.

17.8 Differential Diagnosis

- Posterior ischemic optic neuropathy
- Optic neuritis
- Nonorganic vision loss
- Pre-/intra-/subretinal hemorrhage
- Choroidal rupture
- Commotio retinae

17.9 Treatment

It remains controversial and targets the recovery of secondary axonal damage mechanisms. The

main current options are corticosteroids and surgical decompression, both non-risk-free and without clear evidence in trials.

Moreover, 40–60% of TON have **spontaneous recovery,** excluding severe cases [7–9].

17.10 Corticosteroids

There is no standardized dose. It is based on small retrospective series, anecdotal cases, and extrapolations from studies on brain and spinal trauma.

The **doses** used may be moderate (60–100 mg oral prednisone), high (1 g/day intravenous methylprednisolone), or megadoses (30 mg/kg IV followed by 5.4 mg/kg/h for 24 h) [10–12].

The **NASCIS 2** (National Acute Spinal Cord Injury Study) compared placebo, naloxone, and methylprednisolone, and only showed some superiority of the latter if administered within the first 8 h after trauma: significant improvement in spinal cord motor and sensory function. However, the design of this study showed numerous gaps that detract from its conclusions [13–15].

The **CRASH** (Corticosteroid Randomization After Significant Head Injury) study, which used steroid megadoses, concluded that they should not be routinely used as the treated group had higher mortality than untreated [16, 17].

17.11 Surgical Treatment

Decompressive optical channel surgery is the most commonly used procedure. It can be performed with endoscopic techniques—less invasive—or extracranial. One of the drawbacks is that the general condition of the patient sometimes forces to differ surgery so it could lose effectiveness. Some of the risks of the procedure are CSF leaks and meningitis; both have decreased with the use of endoscopic techniques: they allow good visualization of the orbital vertex and facilitate a faster recovery with better aesthetic results. It is discussed whether it should be performed after 48 h of corticosteroid, so there is less edema, or immediately, without studies allowing a conclusion to be drawn [18, 19].

Some authors recommend it when bone fragments exist in the optical channel, while others argue that their presence already implies that there may have been direct, difficult-to-recover axonal injury, even if the fragment is removed. The IONTS study endorses this view.

The **fenestration of ON** is another reported surgical procedure. Indicated in hematomas of the optic sheath, in N2 portion. A correct preoperative MRI evaluation is vital in these cases, which requires the exact location of the hematoma, because there is no evidence of its success in interstitial hematoma.

In patients with depressed lateral wall fractures or orbital subperiostic hematomas that compress the ON, the best option seems to be **to perform an Orbitotomy,** which allows the reduction of the fracture or the evacuation of the hematoma respectively. On the other hand, if an orbital hemorrhage is demonstrated that compromises the function of the nerve, the immediate realization of **a Cantotomy that allows** the expansion of the orbital content is a priority. If this does not provide sufficient relief, **Decompressive Orbital Surgery should be performed.**

The **IONTS** (International Optic Nerve Trauma Study 1999), designed specifically to evaluate the treatment of TON, compared observation to decompressive surgery and corticosteroids, but its methodological defects were numerous. The end result was that there was no significant difference between the visual recovery of the three groups. That is, between treating and not treating independently of the type of treatment.

TONTT (Traumatic Optic Neuropathy Treatment Trial 2017), compares the use of **erythropoietin** (EPO) vs corticosteroid treatment vs observation. There were no significant differences in the final VA between the 3 groups and, it should be noted, that the 3 groups showed significant improvement. Risk factors for poorer recovery were the initial VA of Light Perception and Late Treatment (after 3 days) [20–23].

From the evidence presented, we conclude that no therapy of those employed **to date in TON can be considered superior to the rest, even compared to mere observation** of the

patient. The decision of using or not steroids will depend on the evaluation of each particular case, with the most widespread pattern being 1 g of methylprednisolone for 3 days followed by oral prednisone in the following days. As well as the possibility of combining medical and surgical treatment, which is suggested in patients who initially have marked VA commitment.

A recent study "Surgical Decompression or Corticosteroid Treatment of Indirect Traumatic Optic Neuropathy: A Randomized Controlled Trial" (Ann Plast Surg 2020) would confirm this trend. In this trial, all patients (30 enlisted patients) were treated: 12 with surgery and 18 with corticoids, without finding significant differences in outcome between the two groups except in cases of initial VA of Light Perception or Amaurosis that would benefit primarily from surgery. Other recent studies with neuroprotective factors such as amniotic membrane derivatives have shown promising results, but do not yet have clinical application and are still in the experimental stage.

17.11.1 Prognosis

- It is poor in **direct lesions** where there is partial or total damage from the optic nerve.
- In **indirect lesions,** a percentage of spontaneous recovery—40 to 60%—is observed. It does not appear to be any significant difference to date between treating and not treating, although it will depend on individual consideration in each case. For example, the presence of large edema justifies the use of corticosteroids, or an important orbital hematoma, surgery.

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