Traumatic optic neuropathy – our experience

Bachi T. Hathiram · Vicky S. Khattar · Harshal P. Sonawane · Parag J. Watve

Abstract A great deal of controversy surrounds the physiology and management of traumatic optic neuropathy. Needless to say, it has formed the topic of much debate in the past, especially with regard to its surgical management. With the advances in sinus endoscopic procedures, and their extended applications to the orbit and optic nerve, endoscopic optic nerve decompression offers a very good chance for salvaging vision in patients with traumatic optic neuropathy. However, there is no definite protocol laid down in the world literature for this condition, owing partially to the fact that a majority of such cases are not amenable to surgery within the critical period, due to the coexisting morbidities of head injury. There is also much controversy regarding medical versus surgical management of traumatic optic neuropathy. We present here our experience with this condition, and outline the management protocol followed.

Keywords Traumatic optic neuropathy · Optic nerve decompression

Introduction

Traumatic optic nerve damage after craniofacial injury was first described by Hippocrates [1]. Indirect damage to the optic nerve is the most common form of traumatic optic neuropathy, occurring in about 0.5–5.0% of all cases of closed head trauma [2, 3]. Patients usually suffer craniofacial trauma but occasionally mild orbital or eye injury [4, 5]. Traumatic optic neuropathy can occur following an innocent ipsilateral injury over the superior temporal orbital rim and is characterized by vision loss without external or internal ophthalmic evidences of injury to the eye and its nerve [6]. In many cases, due to the coexistent head injury and its associated comorbidities, the visual status may not be amenable for assessment. In fact, major brain injury occurs in 40–72% of patients with traumatic optic neuropathy, the management of which takes precedence [7]. This may often lead to a delay in diagnosis and subsequent timely treatment for a potentially reversible visual loss. Patients with head injury who are suspected to have a coexistent optic nerve trauma (which may be indicated by contusions around the eye, an afferent papillary defect, with corresponding fundoscopic changes of disc edema and vascular congestion, or an actual complaint of loss of vision in one/both eyes) warrants urgent radiological investigations. While a high resolution CT scan of the paranasal sinuses and orbit would reveal any obvious fracture, hemosinus and coexisting trauma such as fractures of the skull base, it would also serve as a road map for surgery. An MRI would be helpful in delineating the integrity of the optic nerve, and also help in ruling out nerve sheath hematomas. Usually a clinical suspicion coupled with positive radiological findings may provide enough grounds for surgical intervention; nevertheless, there are certain other investigations which may be performed when in doubt.

B. T. Hathiram · V. S. Khattar · H. P. Sonawane · P. J. Watve
Department of ENT and Head and Neck Surgery,
T. N. Medical College and B. Y. L. Nair Charitable Hospital,
Mumbai Central, Mumbai - 400 008, Maharashtra, India

B. T. Hathiram (✉)
E-mail: bachi.hathiram@rediffmail.com

Pathophysiology

The part of the optic nerve most vulnerable to blunt trauma of the head is the intra-canalicular segment, which by virtue of its bony course is vulnerable to the fractures and compressive-elastic forces of its surrounding bone, which also being unyielding, allows for no space for inflammatory expansion or hemorrhage.

Optic neuropathy following accidental trauma usually results from two distinct mechanisms: a primary injury as a result of a direct contusive force on the optic canal and nerve, or an elastic deformation of the sphenoid sinus with a transfer of force to the intra-canalicular portion of the optic nerve disrupting its axons and blood vessels [7], which may result in compression of the nerve by bony fragments or a sheath hematoma [8], and if untreated may succumb to a secondary ischemia and continued axon loss due to the swelling of the nerve within its sheath and bony canal, compressing its blood supply [7, 9].

Iatrogenic injury of the optic nerve may occur due to a variety of factors which include; anatomic variations of the course of the nerve such as the type III/IV optic nerve or a nerve coursing through a dominant sphenoid sinus of the opposite side (Figs. 1–3), dehiscent bony canal, erosion of the bony canal due to some disease process, excessive hemorrhage impeding visibility during surgery, etc. Most of these problems can be averted by a thorough study of the CT scan prior to surgery and if required, even review during surgery.

Investigations

Certain specific investigations are warranted in a case of traumatic optic neuropathy, and their principles and indications are briefly outlined below:

- **Afferent pupillary defect:** an absolute or relative afferent pupillary defect indicates that vision is being compromised. It can be tested by shining a bright focused light for a few seconds on the ‘unaffected’ eye, in a dark room, with the patient looking at a distance, and noting the pupillary response, which is normally characterized by miosis. Following this, the light is shone on the ‘affected’ eye, and the pupillary response is compared to its ‘normal’ counterpart. Features sought after are a lack of briskness/sluggish miosis as compared to the normal side, a partial or complete absence of miosis. This usually indicates a defect in the afferent pupillary pathway, and has been termed as a ‘Marcus Gunn’ pupil. It is best interpreted along with fundoscopic and radiologic signs. It may also be elicited subjectively, by asking the patient to compare the difference in the brightness perceived between the unaffected and affected eyes.

Fig. 1 Coronal plain high resolution CT scan of the paranasal sinuses showing a type III optic nerve

Fig. 2 Coronal plain high resolution CT scan of the paranasal sinuses showing a type IV optic nerve

- **Fundoscopy examination:** various appearances can manifest in traumatic optic neuropathy namely, disc edema, congestion of vessels, disc pallor, etc. but the most common presentation is usually a normal looking disc, especially in the early stages.
- **Color vision:** loss of color vision, namely red color vision – patients with optic neuropathy often have red

Fig. 3 Intraoperative endoscopic image showing the left optic nerve coursing through a dominant right sphenoid sinus

color desaturation and a positive response would be that the red color looks “faded”, “pink” or “washed out” [10].

- Visual field defects: almost any type of field defect can be seen in cases of traumatic optic neuropathy, but commonly arcuate, central or hemianopic field defects may be seen [10].
- Visual evoked potentials (VEPs): these represent the cortical response to light stimulus, and may be useful in cases of patients with delayed optic neuropathies, wherein the decision to intervene may depend upon the VEP response.
- In patients who present late, with suspected optic atrophy, ‘slit-like’ or ‘wedge-shaped’ defect can be seen in the prepapillary nerve fiber layer, which may be seen with either a red-free ophthalmoscope, or by optical coherence tomography (OCT). OCT a newer technique that uses low coherence light to penetrate tissue and a camera to analyze the reflected image. It performs circular scans around the optic nerve head to analyze the peripapillary nerve fiber layer, and may be used in the follow-up for patients with traumatic optic neuropathy [10].
- CT scan: a high resolution CT scan of the paranasal sinuses and orbit, with windows taken at a distance of 1 mm, besides providing a ‘road-map’ for surgery as this is of utmost importance in endoscopic surgery (especially helping to detect anatomic variations), also helps to reveal any fractures of the orbital apex and optic canal, with/without impinging fracture fragments. It may also reveal any coexisting injury to the skull base, and other structures in the vicinity of the sphenoid sinuses, which may warrant additional precautions and/or simultaneous surgical interventions, for example cerebrospinal fluid (CSF) leaks. It can also

detect any anatomical variations which may impede surgical access to the optic nerve. Usually brain scans should also be done to rule out coexisting brain injury, since the intra-canalicular portion of the optic nerve is within the skull base.

- MRI scan: an MRI is useful to detect optic atrophy, the integrity of the nerve, as well as to rule out nerve sheath hematomas.

The decision to operate or conserve - management protocols

Much controversy clouds the definitive management of traumatic optic neuropathy. Notwithstanding the numerous anecdotal and meta-analytical reports in world literature, stating visual recovery following surgery with/without steroid therapy, various centers-to-date are practising differing protocols.

Carta et al. [11] have identified four negative prognostic factors, which may help in determining the eventual visual prognosis in cases of traumatic optic neuropathy following head trauma, which include:

- Presence of blood within the posterior ethmoid cells
- Age over 40 years
- Loss of consciousness associated with traumatic optic neuropathy
- Absence of recovery of visual acuity after 48 hours of steroid therapy.

Comparative data from the International Optic Nerve Trauma Study (IONTS) group show that neither corticosteroid therapy nor optic canal decompression are the gold standard for treatment of traumatic optic neuropathy. However, on the other hand, Cook et al. [12] in a meta-analysis, reported that recovery of vision in patients treated with mega-dosage steroids or surgical decompression of the optic canal was significantly better than recovery in patients receiving no treatment. A step further, it was shown that high-dose steroids may even be harmful to the optic nerve if started 8 hours after the injury [13]. And again, there are anecdotal reports of a beneficial outcome of surgery in cases of optic canal fracture and optic nerve sheath hematoma [14–16].

Thus, with numerous conflicting reports on the management of traumatic optic neuropathy, there is little world consensus on the optimal management of this condition. Keeping in mind the above, we have devised a management protocol for the same.

The role of medical therapy

The rationale for the use of mega-dose methylprednisolone for traumatic optic neuropathy is based on the improvement

seen with the same in cases with acute spinal injuries; in fact, similar doses have also been recommended, i.e. a loading dose of 30 mg/kg given intravenously, followed by a maintenance dose of 5.4 mg/kg/hr, with monitoring of visual acuity [12]. The timing is crucial for starting this therapy, and there have been numerous reports in literature as regards the importance of initiating the therapy as soon as possible. There have also been reports of mega-dose steroids causing deleterious effects to the visual outcome, if started after 8 hours of trauma [13]. Notwithstanding these anecdotal mentions in literature, most centers today have incorporated this therapy as the initial definitive management; the subsequent visual prognosis being the deciding factor for further surgical intervention versus continuation of mega-dose steroid therapy. The usual cut-off period after the initiation of mega-dose steroids at which the decision for surgical decompression may be undertaken is 48 hours [8]; thus failure of improvement or even worsening of the visual status after initiation of mega-dose steroid therapy for 48 hours, would warrant surgical optic nerve decompression.

The common known side-effects of mega-dose steroid therapy such as transient hyperglycemia and hypertension with cardiac changes, besides the other systemic long- and short-term effects should be kept in mind with the appropriate monitoring of required parameters.

It may be kept in mind that the initiation of steroid therapy would form the initial line of therapy, regardless of the mechanism of traumatic optic neuropathy. This may also buy time for radiological investigations to be undertaken, besides being helpful in reducing the primary and secondary ischemia of the optic nerve due to edema within its bony canal.

Should the radiological investigations suggest an obvious fracture with fragment(s) impinging on the optic nerve, the 48 hours wait-period should be waived and an immediate surgical optic nerve decompression be performed. It is assumed that all cases in discussion would have a severe visual compromise to start with (<6/60 at presentation).

There is again no consensus regarding the duration of mega-dose steroid therapy, but an average minimum of 3–5 days is usually warranted, which may be followed up with a lower maintenance dose for 7–10 days, with continuous monitoring of blood sugar levels and the cardiac status, besides the usual adjunctive measures such as calcium supplements, etc.

Optic nerve decompression

Having said much about the followers and non-believers of this potentially gratifying surgical procedure, there are certain definitive indications for traumatic optic nerve decompression, namely:

- Radiologically evident bony fracture fragment impinging on the intra-canalicular portion of the optic nerve in the lateral wall of the related sphenoid sinus (Fig. 4), or an optic nerve sheath hematoma (as seen on MRI), in a patient with traumatic optic neuropathy with a vision of <6/60 at presentation
- Failure to improve/deterioration of vision after 48 hours of mega-dose steroid therapy in a patient with traumatic optic neuropathy with vision <6/60 at presentation, with no obvious radiological evidence of compromise of the volume of the optic canal by a hematoma or fracture fragment impingement. This may include any probable canal injury [8] in patients with a similar clinicoradiological profile, indicated by the presence of fluid levels in the posterior ethmoid and sphenoid sinuses, and/or the presence of fractures of the ethmoids, orbital apex and sphenoid.

The erstwhile external and transcranial approaches for optic nerve decompression have given way to a new gold standard – endoscopic optic nerve decompression. There are many advantages of the latter. In the hands of an experienced endoscopic sinus surgeon, this procedure is a relatively safe operation (Figs. 5–8), with a greatly reduced morbidity, and has the potential to improve, and in some cases restore lost vision, especially after blunt trauma.

There are certain potential complications associated with this procedure, some of which may be a consequence of the trauma which may have caused the optic neuropathy in the first place, and some may be due to certain anatomical variations and considerations which should be kept in mind.

Concomitant damage to the base of the skull with a resultant CSF leak may be as a result of the trauma, or

Fig. 4 Coronal plain high resolution CT scan of the paranasal sinuses showing a fracture fragment impinging on the left optic nerve with corresponding sphenoid hemosinus

Fig. 5 Intraoperative endoscopic image of the same patient (Fig. 4) showing the fracture fragment being gently teased off the underlying traumatized left optic nerve with a blunt probe

Fig. 8 Panoramic intraoperative endoscopic view of the same patient (Fig. 4) showing the decompressed left orbital apex and optic nerve

Fig. 6 Intraoperative endoscopic image of the same patient (Fig. 4) showing the fracture fragment being removed with the tip of the forceps

Fig. 7 The fracture fragment after removal – note the sharp ragged edges which resulted in injury to the nerve

iatrogenic (Fig. 9). In any case, once detected should be managed simultaneously endoscopically. There are certain factors which must be kept in mind for the same. Whereas in endoscopic optic nerve decompression, we result in an exposed nerve, and any packing of the nasal cavity, especially in the region of the ethmoid and sphenoid sinuses should be avoided at all costs, in CSF leak repair, we may have to go in for a multilayered closure, with packing to support our closure material, till adhesion formation takes place. Now any pressure on an exposed optic nerve may result in visual compromise; this can sometimes pose a problem. This may be solved in many cases by the use of

Fig. 9 Coronal plain high resolution CT scan of the paranasal sinuses of the same patient (Fig. 4) showing a concomitant fracture of the planum sphenoidale. The patient had a coexisting CSF leak along with a left traumatic optic neuropathy

a ‘bath-plug’ technique using autologous fat (Figs. 10a and 10b), as has been described by Wormald [8]. Some centers also propagate the use of tissue glue to keep the graft (fascia lata, etc.) in place, thus potentially avoiding the need for packing materials and multilayered closure for co-existent CSF fistulae; nevertheless, this has been the subject of much controversy. This is more so true, when the site of CSF leak is very close to the optic nerve, such as in the roof of the sphenoid sinus.

Also, because the sheath of the optic nerve is a direct continuation of the dura mater, there is always a chance of a potential CSF leak following optic nerve decompression, in cases where the sheath has been split so as to release the edema, and also to evacuate any underlying hematoma. While this may theoretically pose a problem, it is rather unlikely, since traumatized optic nerves are usually edematous, and following splitting of their sheath would bulge outwards

and obliterate any such ‘potential space’, thus preventing a CSF leak.

The ophthalmic artery which usually runs through the postero-inferior quadrant of the optic nerve may sometimes run anomalously around the latter’s lower edge, and into the surgical field [17]. The risk of damage to this artery may be minimized by incising the nerve sheath in its upper medial quadrant [18, 19].

Other potential complications may be damage to the medial rectus muscle while incising the periosteum over the orbital apex (since the pad of fat between the medial rectus muscle and the orbital periosteum is very thin in this region), and other known complications of endoscopic sinus surgery in general.

Nevertheless, there are numerous advantages to this procedure: it is a minimally invasive scar-less surgery, avoids open and neurosurgical approaches which are associated with a greater morbidity, allows for an end-on visualization of structures, making accurate instrumentation possible, allows for simultaneous management of CSF leaks, and is very suited for fracture fragments in the medial wall of the optic nerve.

There are certain limitations for endoscopic optic nerve decompression, which include fragments impinging on the lateral aspect of the optic nerve and patients with other cranial and skull base injuries which may then be concomitantly managed via an open neurosurgical approach.

Conclusion

Traumatic optic neuropathy is a potentially reversible condition (partially or completely), if managed on an urgent basis by experienced personnel. Due to lack of sufficient supportive evidence, there is no clear consensus on the management protocol to be followed. Proponents are aplenty for medical versus surgical management, and there also exists a third category of physicians and surgeons who choose to ‘wait and watch’. However, keeping in mind the disastrous consequences of vision loss, it would be a reasonable proposition to offer the patient the best chance of salvaging vision. A combined modality protocol including mega-dose steroid therapy and endoscopic optic nerve decompression probably is one such option.

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Fig. 10a The ‘bath-plug’ being prepared

Fig. 10b Intraoperative endoscopic image showing the ‘bath-plug’ sealing off the defect in the planum sphenoidale causing an immediate cessation of the CSF leak

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