Chapter 8
Optic Nerve Sheath Fenestration in Cancer Patients: Indications and Surgical Technique

Thomas E. Johnson

Abstract Optic nerve sheath fenestration (ONSF) entails cutting a window or making linear fenestrations in the retrobulbar optic nerve sheath, which releases pressure and often allows stabilization or improvement of vision. Indications for ONSF include visual loss due to pseudotumor cerebri, optic nerve sheath hemorrhage, dural sinus thrombosis, subdural hematoma, intradural arteriovenous malformation, arachnoiditis with increased intracranial pressure, and cryptococcal meningitis with papilledema due to AIDS. Indications for ONSF in cancer patients are not well established, but a few case reports have shown success of ONSF in patients with perineural metastasis of breast cancer, increased intracranial pressure with papilledema due to a brain tumor, lymphomatous infiltration of the optic nerve, and optic nerve sheath meningioma. ONSF can be performed with a medial orbitotomy approach with disinsertion of the medial rectus muscle, a superomedial eyelid crease incision without extraocular muscle disinsertion, a lateral orbitotomy approach with bone removal, or a lateral canthotomy incision without bone removal. ONSF is considered relatively safe when performed carefully; serious complications occur in about 1% of patients.

8.1 Introduction

The surgical technique of optic nerve sheath fenestration (ONSF) is used most often in the treatment of progressive optic neuropathy in patients with idiopathic increased intracranial pressure (pseudotumor cerebri) in whom medical management has failed. Pseudotumor cerebri is characterized by elevated intracranial pressure and papilledema (Fig. 8.1) in the context of normal neuroimaging studies and cerebrospinal fluid analysis and is a diagnosis of exclusion. Typically, affected patients

T.E. Johnson (✉)
Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL, USA
e-mail: tjohnson@med.miami.edu

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Fig. 8.1 Photograph of optic nerve head swelling in a patient with increased intracranial pressure

are obese females of child-bearing age who present with headache and visual obscuration. In severe cases, patients may report diplopia secondary to cranial nerve VI palsy. The increased nerve sheath pressure can result in progressive visual field loss and even blindness in advanced cases.

Medical treatment includes encouragement of weight loss and oral administration of a carbonic anhydrase inhibitor (e.g., acetazolamide). Furosemide and systemic corticosteroids may also be used. If maximal medical therapy fails or if patients develop intolerance to the medications, surgery is often required. Options include neurosurgical cerebrospinal fluid shunting, using either a ventriculoperitoneal or a lumboperitoneal shunt, and ONSF.

Cutting a window or making linear fenestrations in the retrobulbar optic nerve sheath releases pressure and often allows stabilization or improvement of vision. The mechanism of action is unknown but most likely relates to filtration resulting from successful creation of a meningeal space–orbital fistula [1]. Hamad and coauthors demonstrated cyst-like structures contiguous with the fenestration sites after ONSF on magnetic resonance imaging and echography. They concluded that the procedure creates a filtration apparatus that controls the pressure in the subarachnoid space surrounding the intraorbital segment of the optic nerve [2]. Many patients—in some series, over 50%—experience bilateral resolution of papilledema after unilateral ONSF [3, 4]. ONSF stabilizes or improves vision in the majority of patients with pseudotumor cerebri and visual loss, but the procedure may fail any time after the surgery, and patients need to be followed up routinely [5].

8.2 Indications

In addition to pseudotumor cerebri and visual loss, indications for ONSF include visual loss secondary to optic nerve sheath hemorrhage, dural sinus thrombosis, subdural hematoma, intradural arteriovenous malformation, and arachnoiditis with increased intracranial pressure. Dural sinus occlusion can produce a clinical picture similar to pseudotumor cerebri with elevated intracranial pressure and papilledema [6]. Additionally, ONSF has been employed in the treatment of visual loss in patients with AIDS and cryptococcal meningitis with papilledema [7]. ONSF was not found to be helpful and was found to be possibly harmful in patients with nonarteritic ischemic optic neuropathy [8].

The indications for ONSF in cancer patients are not well established, but a few case reports have shown success of ONSF in patients with neoplastic disease, including patients with perineural metastasis of breast cancer [9], lymphomatous infiltration of the optic nerve [10, 11], increased intracranial pressure with papilledema due to a brain tumor [12], and optic nerve sheath meningioma [13].

8.3 Surgical Techniques

The first optic nerve sheath decompression was described by De Wecker in 1872 [14]. Since then, many different surgical techniques have been described. The optic nerve can be approached through a medial orbitotomy with disinsertion of the medial rectus muscle, a superomedial eyelid crease incision without extraocular muscle disinsertion, a lateral orbitotomy with bone removal, and a lateral canthotomy incision without bone removal. The nerve sheath can be opened either by excising a window of dura and arachnoid or by making multiple linear incisions in the nerve sheath. Most cases are performed under general anesthesia. Preoperative testing usually includes visual acuity measurement, color vision assessment, visual field evaluation using quantitative perimetry, and fundus examination with attention to the optic disks. Most patients require a magnetic resonance imaging scan and/or a computed tomography scan of the brain and orbits to rule out hydrocephalus, space-occupying masses, or vascular malformations that can also cause papilledema. A magnetic resonance venogram is helpful in evaluating patients suspected of having a venous sinus thrombosis.

Instrumentation for ONSF includes surgical loupes and/or an operating microscope, Freer periosteal elevators, ribbon malleable retractors, neurosurgical microforceps, bayonet neurosurgical scissors (e.g., Yasargil bayonet scissors), neurosurgical cottonoids, and a headlight. Four main surgical approaches have been described for ONSF.

8.3.1 Medial Orbitotomy Approach

For the medial orbitotomy approach, a 270° medial conjunctival peritomy is made using Westcott scissors. The quadrants between the medial rectus and the superior rectus and between the medial rectus and the inferior rectus are cleared with Stevens scissors. The medial rectus muscle is isolated on a muscle hook, and a double-armed 5-0 polyglactin (Vicryl) suture is passed through the muscle insertion and locked on each end. The medial rectus is disinserted, allowing access to the medial orbital space. Gentle dissection is carried out with blunt Freer elevators or thin ribbon malleable retractors. Slightly dampened neurosurgical cottonoids are used to gently retract the orbital fat. Attention is carefully paid to the pupil, and all pressure is removed if pupillary dilation occurs. The nerve sheath is exposed just behind the globe, where it is most bulbous (Fig. 8.2). Often, there is a fine vascular plexus on the epidural sheath, and these vessels should be avoided when a nerve sheath window is excised. Either an adjacent area of the sheath can be excised or the vessels can be gently moved using the blunt end of a Freer elevator. A neurosurgical microforceps is used to grasp the dural sheath just behind the globe, and a window of approximately 3 mm × 5 mm is cut using bayonet microsurgical scissors. Care is taken to cut through both the dural and the arachnoid sheaths to allow egress of cerebrospinal fluid, and the surgeon should carefully avoid touching the optic nerve within the sheath. A gush of clear cerebrospinal fluid is usually noted when the sheath is adequately opened. After careful inspection to assure meticulous hemostasis, the medial rectus muscle is reattached using the 5-0 Vicryl sutures, and the conjunctiva is closed using 6-0 fast-absorbing plain sutures. An antibiotic–steroid ophthalmic ointment is applied to the eye, and an eye pad is placed over the closed eyelids. No pressure is applied to the eye. Postoperative icepacks are helpful to reduce swelling. The ointment is applied three times per day for a week, and the patient is instructed to avoid any blood thinners, such as aspirin and nonsteroidal anti-inflammatory medications, as well as herbal medications for 1 week.

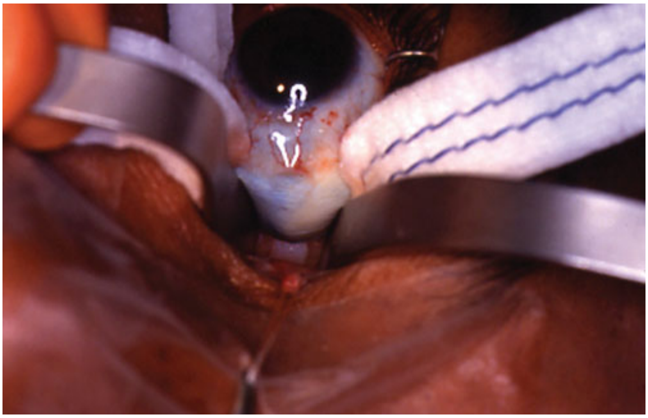

Fig. 8.2 Medial orbitotomy approach to ONSF with disinsertion of the medial rectus muscle and use of neurosurgical cottonoids and malleable retractors

8.3.2 Medial Eyelid Crease Approach

Pelton and Patel [15] described using a medial eyelid crease approach to ONSF without the need for disinsertion of a rectus muscle. A 1-cm eyelid-crease incision is made over the medial third of the upper eyelid, and the incision is carried through skin and orbicularis muscle. Scissors are used to open the orbital septum, and blunt dissection is used to open fascial septi in the superomedial compartment. The medial intraconal space is exposed using gentle retraction with Sewall orbital retractors, and the standard fenestration instruments are used to open a rectangular window in the nerve sheath. Stated advantages include ease of dissection, short incision-to-nerve distance, and angle of approach to the optic nerve.

8.3.3 Lateral Orbitotomy Approach

Tse and coauthors [16] described using a traditional lateral orbitotomy approach with elevation of a bone flap to perform ONSF. They used an operating microscope after adequate exposure of the nerve sheath was accomplished. The lateral rectus muscle was isolated from the bulbar conjunctiva with a 4-0 silk suture, and the globe was adducted in order to move the optic nerve closer to the lateral wall. A window was excised from the bulbous lateral aspect of the retrobulbar nerve sheath. Stated advantages of this approach are that it allows a direct, perpendicular view of the optic nerve using an approach well known to the orbital surgeon and does not require the disinsertion of a rectus muscle. Disadvantages include the need for general anesthesia, longer operating time than the other approaches to ONSF, and greater risk of damage to the ciliary ganglion with resultant pupillary dilation.

8.3.4 Lateral Canthotomy Approach

Kersten and Kulwin [12] described a technique of approaching the optic nerve laterally without elevating a bone flap. A lateral canthotomy and inferior cantholysis exposes the lateral orbital rim. A 4-0 silk suture is placed under the lateral rectus muscle insertion from the bulbar conjunctiva to adduct the globe and bring the nerve closer to the lateral wall. The periosteum on the rim is cut, and the periorbita is elevated 2 cm posterior to the rim. After opening of the periorbita, Sewell retractors are used to retract the lacrimal gland superiorly and the lateral rectus inferiorly. Orbital dissection is gently carried out to expose the nerve, and a fenestration is performed using the operating microscope and standard fenestration instruments. Reported advantages of this technique include ease of operation, as with the medial eyelid crease approach, and a more direct perpendicular view of the nerve, as with the standard lateral orbitotomy approach with bone removal.

8.4 Possible Indications for ONSF in Cancer Patients

As mentioned earlier in the chapter, information about possible indications for ONSF in cancer patients comes mainly from case reports.

8.4.1 Metastatic Breast Cancer

Gasperini and coauthors [9] reported a 36-year-old woman with bilateral anterior optic neuropathy and orbital and central nervous system metastasis of breast cancer. She had progressive bilateral visual loss and bilateral optic disc swelling. ONSF on the left optic nerve by the medial eyelid crease approach resulted in bilateral visual improvement and resolution of disc edema in both eyes. The etiology of bilateral optic neuropathy in this patient was thought to be due to papilledema caused by perineural infiltration by the metastatic breast cancer. The authors concluded that ONSF should be considered as a treatment option for optic neuropathy caused by perineural or intrasheath metastasis [9].

8.4.2 Lymphomatous Optic Neuropathy Diagnosed by Optic Nerve Biopsy

ONSF has been used to diagnose cancer. Dayan and coauthors [10] diagnosed a 74-year-old woman with lymphomatous optic neuropathy by performing an optic nerve biopsy using an ONSF approach. Gross examination of the optic nerve sheath was not diagnostic, but the optic nerve substance showed infiltration with a low-grade B-cell non-Hodgkin lymphoma. The patient had temporary visual deterioration after biopsy but later showed improvement to at least prebiopsy vision. The authors concluded that optic nerve biopsy using an ONSF approach should be considered in enigmatic cases with signs or symptoms suggestive of optic neuropathy even when visual function remains [10].

Kitzmann and coauthors [11] reported a 39-year-old man with T-cell non-Hodgkin lymphoma and bilateral optic nerve infiltration. ONSF was performed, and pathologic examination showed only meningeal dural tissue with no lymphocytes. After dramatic initial visual improvement, the patient's vision decreased to no light perception by postoperative day 17. The authors concluded that ONSF does not appear to be helpful in restoring vision in patients with lymphomatous infiltration of the optic nerve [11].

8.4.3 Adjuvant Therapy in Optic Nerve Sheath Meningioma

Turbin and coauthors used ONSF as adjuvant therapy in two patients with optic nerve sheath meningioma and progressive visual loss. One had received fractionated stereotactic radiotherapy before surgery, and the second patient received radiotherapy after ONSF. In both patients, the visual loss was thought to be due to nerve compression within a tight sheath. Both patients experienced improvement of vision and resolution of disc edema after ONSF. The authors concluded that in selected patients with optic nerve sheath meningioma, severe disc edema, rapid visual loss, and few treatment options, ONSF may have an important but restricted role as an adjuvant to radiotherapy [13].

8.4.4 Papilledema Associated with Brain Tumors

Kersten and Kulwin reported that one of the patients in their series of patients who underwent ONSF had papilledema and visual loss due to an intracranial glioblastoma. That patient's vision improved after ONSF performed using a lateral canthotomy approach. Disk edema cleared in both eyes after unilateral ONSF [12].

8.4.5 Radiation-Induced Optic Neuropathy

Radiation-induced optic neuropathy is a well-recognized complication of external-beam radiotherapy and can result in progressive unilateral or bilateral visual loss months to years after radiotherapy has been completed. Radiation can cause edema and increased perineural pressure in the optic nerve. Mohamed and coauthors treated three patients with pending anterior radiation-induced optic neuropathy with ONSF by a medial orbitotomy with disinsertion of the medial rectus, and all three patients experienced resolution of edema and restoration of vision [17]. ONSF was used proactively in these three cases of impending radiation-induced optic neuropathy to relieve the swelling and increased pressure within the confines of the nerve sheath [17].

8.5 Complications of ONSF

ONSF is considered a relatively safe surgery when performed carefully; serious complications occur in about 1% of patients [18, 19]. Possible complications include partial or complete visual loss due to central retinal artery occlusion, branch retinal artery occlusion, traumatic optic neuropathy due to excessive traction on the nerve, and hemorrhage within the optic nerve sheath. Blindness has been reported in about 1% of ONSF cases. Ocular motility disorders can be transitory or permanent and may be due to medial or lateral rectus muscle damage or a lost muscle. Anisocoria can occur because of injury to the ciliary ganglion or to the ciliary nerves lying along the lateral optic nerve, especially when the nerve is approached from the lateral aspect. Complete blindness with slow recovery of vision has been reported following ONSF performed by the medial approach. The mechanism was thought to be sustained rotational traction on an already compromised optic nerve with resulting axonal demyelination from stretch injury, followed by remyelination of affected axons [20].

8.6 Future Research

Joos and coauthors reported the experimental use of an endoscopic surgical approach to ONSF with the use of the free electron laser in a goat model [21]. Their work creates the potential for performing minimally invasive ONSF in the future [21].

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