Ophthalmology Research

Is Papilledema Really An Increasing Neuro-Ophthalmological Condition Today?

Cuneyt Karaarslan*

*Correspondence:

World Eye Hospital, Adana Branch, Turkey.

Cuneyt Karaarslan, World Eye Hospital, Adana Branch, Turkey, Tel: +90 533 7317273, Fax: +90 322 4571914.

Received: 14 Jan 2024; Accepted: 21 Feb 2024; Published: 27 Feb 2024

Citation: Cuneyt Karaarslan. Is Papilledema Really An Increasing Neuro-Ophthalmological Condition Today?. Ophthalmol Res. 2024; 7(1); 1-7.

ABSTRACT

The optic nerve is the name of the visual nerve that takes the longest path within the skull, formed by the axonal extensions of specialized nerve cells of the retina. Raised edematous swelling in which the borders of the optic nerve head can be erased is called papilledema. Although the cause of papilledema cannot often be determined exactly, the real cause can be revealed with a detailed and careful examination. Unlike true papilledema, there is no increase in intracranial pressure in pseudo-papilledema, and therefore diagnosis and detection are more difficult. In the diagnosis of papilledema, it is very important to detect edematous swelling of the optic nerve head directly with the ophthalmoscope. However, a more detailed examination and anamnesis are often required for correct diagnosis. While there are many neurological and ophthalmic complaints and findings in true papilledema, neither applies to pseudo-papilledema. It is essential to distinguish all these conditions well for a successful and sequefree treatment.

Keywords

Papilledema, Pseudopapilledema, Optic nerve, Vision loss, Neuro Ophthalmology.

Introduction

Many patients with headaches, fuzziness, and various neurological and ophthalmological complaints, which were thought to be due to COVID-19, empirical conventional drug treatments, or vaccines, applied to our clinic during the pandemic period because of the preliminary diagnosis or suspicion of papilledema. When the necessary and detailed examinations were carried out, it was found that these patients actually had pseudo-papilledema and/ or relatively innocent diseases of the optic disc, which were congenital or acquired and were of lesser importance, or at least were not likely to result in blindness or had no risk of mortality. This situation, especially during the pandemic period, was considered to be associated with insufficient, incomplete, and hastily obtained patient records, the inability to perform a close ophthalmological examination, and the lack of further examination opportunities in emergency clinics. The vital importance of true papilledema and its differential diagnosis (because of the risk of blindness and

Ophthalmol Res, 2024

mortality) must be emphasized in this respect.

Although there is no current increase in the frequency of papilledema today, it may be generally misdiagnosed by careless examinations. Such a situation may be a matter of the recent COVID19 pandemic due to the limited examinations in pandemic rules. Although many cases of papilledema have been reported that are assumed to be related to the Coronavirus pandemic, this situation is not very clear yet. Papilledema can be associated with severe COVID-19 infection, but it may also be mistaken for pseudo-papilledema or optic disc edema due to careless and rushed examinations, especially during such outbreaks. Making an accurate differential diagnosis of papilledema, pseudo-papilledema, and optic disc edema is very crucial to prevent the patient from expensive invasive procedures and especially from blindness.

Optic Nerve

The optic nerve is the second one of the twelve cranial nerves and it is responsible for transmitting all visual stimuli to the central nervous system. It is made up of over one million myelinated axons of the retinal ganglion cells that have only sensory fiber. The length of the optic nerve is approximately 35-55mm and there are four compartments in the optic nerve (intra-ocular, intra-orbital, intra-canalicular, and intracranial). The intra-ocular part is also known as the optic nerve head and is located in the back, right after the eye with a length of about 1mm. the intra-orbital part is within the orbital eye socket with a length of about 25mm.



Figure 1: Optic nerve anatomy.

The intra-canalicular part is within an osseous canal with a length of about 4-10mm. the intracranial part is located in the cranial cavity, through the optic chiasm where there is a partial decussation (crossing) of temporal visual field fibers with a length of about 10mm. The optic nerve transmits all visual data and information including brightness, perception in darkness, color perception, contrast sensitivity, and visual acuity from the retina to the occipital cortex in the brain. Any damage to the optic nerve can lead to vision loss unilaterally or in both eyes and optic nerve disease can cause various visual symptoms and signs depending on the underlying condition. The treatment of optic nerve disorders depends on the underlying cause and may include medications, surgery, or other interventions.

Which Optic Nerve Diseases are Confused with True Papilledema and What are the Clinical Durations of these Diseases?

The following types of optic nerve disorders can affect vision.

- 1. **Optic neuritis:** Inflammation of the optic nerve, which can be caused by infections or immune response-related diseases such as multiple sclerosis.
- 2. **Glaucoma:** In fact, this is a group of disorders that can cause serious optic nerve damage due to increased intra-ocular pressure.
- 3. **Papilledema:** There is a swelling of both optic discs due to increased intracranial pressure.
- 4. **Pseudo-papilledema:** This clinical condition can be defined as an anomalous elevation of one or both optic discs without a raised intracranial pressure.

Optic nerve atrophy: The optic nerve is highly sensitive to poor blood flow into the eye. Certain systemic vascular disorders, ocular, orbital, or cranial traumatic events, or exposure to some neurotoxic substances can cause optic nerve atrophy due to the lack of nourishment.

Optic nerve head drusen: The deposits of protein and calcium salts in the optic nerve create these structures.

Optic nerve pit: An abnormality of the optic nerve head that causes loss or decrease in connection between the retina and the optic nerve congenitally.

Neuromyelitis optica: An autoimmune disease that can cause an inflammation of the optic nerve and spinal cord.

Coloboma of the optic nerve: There is a defect in the optic disc on account of the maldevelopment.

Idiopathic intracranial hypertension: An increased pressure within the skull causes compression and damage to the optic nerve. Septo-optic dysplasia: A genetic disorder that affects the development of the optic nerve and the chiasm, leading to vision loss.

Toxic amblyopia (nutritional amblyopia): Vision loss caused by a lack of nutrients, such as vitamin A.

Diagnosis

The exact diagnosis of optic nerve disorders can be achieved with a well-combined and detailed medical history, ophthalmic examination, and specific neurologic and ophthalmologic tests. The methodology that is missing or needs to be clarified must be as follows.

- 1. **Examination:** A routine eye examination includes evaluation of visual acuity, central and peripheral vision, color, and contrast perception.
- 2. **Ophthalmoscopy:** The ophthalmologist examines the structures at the back of the eye through the pupil including the retina and the optic nerve disc.

Optical coherence tomography (OCT-RNFL): This test measures the thickness of the ocular retinal nerve fiber layer. **Visual field test:** This test measures the central and peripheral visual areas.

Magnetic resonance imaging (MRI): Imaging tests such as MRI can help identify structural abnormalities or inflammation in the optic nerve.

Serologic analysis and blood tests: These tests may be performed to identify underlying autoimmune, inflammatory, or infectious conditions that can cause optic nerve disorders. Early diagnosis is highly crucial for optic nerve disorders to prevent permanent damage to visual functions and total blindness.

Is it Papilledema or Not?

Papilledema is the swelling of the optic disc due to increased intracranial pressure (ICP). Papilledema can be caused by brain tumors, meningitis, cerebral venous sinus thrombosis, Idiopathic Intracranial Hypertension (IIH), or some other various intracranial conditions. It is a serious condition that can lead to vision loss and requires prompt medical attention. Especially IIH, as an insidious and common cause of papilledema, has some risk factors such as obesity, recent excessive weight gain, thyroid disease, anemia, polycystic ovarian syndrome, and obstructive sleep apnea.



Figure 2: Normal optic disc and papilledema.

Symptoms and Signs

Even if papilledema may not cause any symptoms in some cases initially (may be discovered during a routine eye examination), it presents commonly along with various symptoms.

- Vision problems, such as blurred or double vision, flickering, or complete loss of vision.
- Headaches, often worse in the morning.
- Nausea and vomiting.
- Hearing vascular noises (pulsatile tinnitus).

As the condition progresses, untreated papilledema can lead to blindness. It is essential to seek medical attention if there are any of the symptoms mentioned above, especially if they worsen or persist.

Papilledema is an important condition characterized by the swelling of both optic discs in the eyes due to increased intracranial pressure (ICP), so we need to find the underlying cause of increased intracranial pressure to prevent further complications. Some possible causes of increased ICP and papilledema include brain tumors, meningitis, cerebral hemorrhage, hydrocephalus, spinal cord lesions, and impaired cerebral sinus drainage. It shall not be forgotten that papilledema can lead to serious vision problems and even blindness if left untreated. Early diagnosis and treatment are crucial for better outcomes. Long-term effects of papilledema can be severe and may include the following signs and symptoms.

- 1. **Visual impairment:** In advanced stages, papilledema may lead to an enlargement of the blind spot in visual field measurements, blurred vision, visual obscurations, or even total loss of vision.
- 2. The timeline of visual impairment associated with papilledema is highly variable, with major deficits arising within weeks in severe cases, but more typically they arise over several months.
- 3. **Nerve fiber dysfunction:** Papilledema can cause nerve fiber dysfunction from swelling, which may result in progressive visual field loss and loss of central visual acuity.
- 4. **Permanent dropout of the nerve fiber layer:** Chronically high ICP can lead to permanent dropout of the nerve fiber layer, which can result in progressive visual field loss and loss of central visual acuity.
- 5. Complications: Untreated papilledema and increased

intracranial pressure may lead to brain damage, stroke, seizures, constant headaches, and even death.

The prognosis for papilledema is related to the chronicity of the disease. Prompt treatment of the underlying cause is crucial to prevent long-term complications and vision loss. Treatment for papilledema primarily focuses on addressing the underlying cause of the increased ICP, which may involve surgery, medication, or other interventions.

Differential Diagnosis Pseudo-Papilledema

Pseudo-papilledema is a significant and anomalous elevation in one or both optic nerve heads. There is no increased intracranial pressure or retinal nerve fiber layer edema in this clinical condition. The most common causes of accompanying clinical manifestations of pseudopapilledema are as follows.

- 1. Optic nerve head drusen.
- 2. Congenital anomalies: Congenitally small optic nerve with a crowded optic nerve head, often seen in hyperopic eyes with shorter axial lengths.
- **3. Myelinated nerve fibers:** In myopic eyes with longer axial lengths, the nerve fiber layer may be partially myelinated, leading to the appearance of pseudo-papilledema.
- 4. **Peripapillary masses:** Tumors such as astrocytic hamartomas can cause pseudopapilledema.
- 5. Vitreopapillary traction: This can also lead to the appearance of pseudo-papilledema.
- 6. Systemic causes: Conditions such as diabetic papillopathy, toxic insult, metabolic or nutritional deficiency, optic perineuritis (caused by infectious or inflammatory conditions), optic neuritis, ischemia, and compression from orbital tumors can contribute to pseudo-papilledema.

The main difference between papilledema and pseudo-papilledema (optic disc edema) lies in their underlying causes. Papilledema specifically refers to optic disc edema caused by increased intracranial pressure (ICP), often indicating a serious medical condition. On the other hand, pseudo-papilledema more broadly refers to the swelling of the nerve fiber layer at the optic nerve head due to various causes such as inflammatory, infiltrative, or compressive optic neuropathies. Papilledema is a critical condition that requires prompt medical attention, as it may be associated with serious health problems such as brain tumors, meningeal infections, cerebrovascular anomalies and hemorrhages, and excessive production of impaired drainage of cerebrospinal fluid.

On the contrary, pseudo-papilledema does not have to be linked with a raised ICP and it may be derived from some other underlying causes. While papilledema specifically denotes optic disc edema due to raised intracranial pressure, optic disc edema in pseudopapilledema is a more general term encompassing a variety of conditions leading to swelling of the optic nerve head.

Optic disc edema and pseudo-papilledema are not the same, but they share some similarities in appearance. While optic disc edema

refers to the swelling of the nerve fibers at the optic nerve head due to infiltrative or compressive reasons, pseudo-papilledema defines an anomalous elevation in one or both optic discs without edema of the retinal nerve fiber layer. Papilledema, on the other hand, is a swelling of the optic disc due to increased intracranial pressure. These conditions can cause the optic disc to appear elevated, but they have different clinical implications and require different approaches. Pseudo-papilledema and optic disc edema can be caused by various factors, including congenital optic disc anomalies, myelinated nerve fibers, and peripapillary masses such as astrocytic hamartomas. In contrast, as a specific type of optic disc edema, papilledema is always caused by raised intracranial pressure. The evaluation of the deterministic differences between papilledema, optic disc edema, and pseudopapilledema is based on a detailed patient examination and medical history. In some confusing cases, additional tests may be necessary to identify the type of elevated appearance in the optic nerve head. Proper diagnosis and management of these conditions are crucial to avoid vision loss, neurological impairment, or death by the conditions mentioned below.

Increased Intracranial Pressure (ICP)

ICP might occur due to conditions such as intracranial spaceoccupying lesions, cerebrospinal fluid (CSF)-producing tumors, obstructions of the arachnoid villi, and Idiopathic Intracranial Hypertension (IIH).

Inflammatory and Infectious Conditions

These can impede the flow of axoplasmic transport in the optic nerve, leading to edema.

Infiltrative Lesions of the Optic Nerve

Tumors or other infiltrative conditions can cause optic disc edema.

Toxic Causes

Acute toxic or nutritional etiologies can also lead to optic disc edema. Hereditary Optic Neuropathies: Certain genetic conditions can be associated with optic disc edema. It is important to note that optic disc edema is a manifestation of several disorders rather than a specific diagnosis, and a thorough clinical investigation is necessary to determine the underlying cause. The main differences between papilledema and pseudo-papilledema are in their etiology and consequences. Pseudo-papilledema can be caused by various factors, including inflammatory or infectious conditions, infiltrative lesions, toxic causes, and hereditary optic neuropathies whereas papilledema is specifically caused by increased intracranial pressure can lead to optic disc edema and optic nerve atrophy.

Clinical Implications

Pseudo-papilledema refers to the swelling of the optic nerve head due to infiltrative or compressive causes and occasionally can lead to some visual and neurological symptoms. Papilledema is also the swelling of the optic disc but it is caused by increased intracranial pressure and can lead to vision loss, neurological impairment, or death due to the underlying disease. It is a more morbid type of optic disc edema and its management focuses on addressing the underlying causes of the increased intracranial pressure.

Prognostic Differences

The prognosis and management of a swelled optic disc depend on its severity, type, and underlying cause. Here, to avoid any misdiagnosis is crucial. In the case of papilledema, the management focuses on treating the underlying cause of the increased intracranial pressure, which may include surgical intervention or medical management of related conditions.

Material - Method and Diagnostic Tools

The causes, implications, prognosis, and treatment of swelled optic nerve conditions are very different. To avoid the possibility of severe complications due to late or misdiagnosis, or expensive and invasive diagnostic procedures, differential diagnosis is crucial. Several diagnostic methods can be useful instead of routine eye examinations. Papilledema may be diagnosed truthfully through a well-planned combination of the following.

- 1. Examination: Including direct and indirect ophthalmoscopy.
- **2. Imaging Tests:** Magnetic resonance imaging (MRI) or computed tomography (CT).
- **3.** Lumbar Puncture or Spinal Tapping: Measuring the pressure of the cerebrospinal fluid.

Table 1: Symptoms and signs.

Features	Papilledema	Pseudo-papilledema
Visual symptoms	May have none early Associated with transient visual obscuration, diplopia May have peripheral visual field constriction or decreased visual acuity in severe cases Associated with sudden vision loss with vascular complications	Depends on etiology Optic neuritis associated with severe vision loss, dyschromotopsia AION may have dense inferior, superior, or central vision loss Visual disturbance may vary with eye movement in severe edema
Visual field defect	May be normal Most common: enlarged blind spot, nasal step, altitudinal visual field loss, constricted visual field	Depends on etiology. May have central scotoma or altitudinal visual field defect. Often abnormal in optic neuritis and AION. Sometimes normal
Other neurological symptoms or syndrome	Commonly associated with pulsatile tinnitus, dizziness, nausea/ vomiting, cranial neuropathies Can be due to idiopathic intracranial hypertension, brain tumor, hemorrhage, meningoencephalitis, venous thrombosis, and trauma	Optic neuritis may be associated with multiple sclerosis or other neurological issues

4. Visual Field Tests: The standard automated perimetry (SAP) is the most commonly used test to evaluate the perception of the visual area and the field screening performance of the optic nerve. To be prone to patient falsity and conspicuous subjectivities of these tests are the major limitations of them.

Visual field tests are the most essential diagnostic tools for optic nerve disorders. According to the severity of the papilledema, the patient may show glaucomatous or glaucoma-like visual field defects. The enlarged blind spot (most common finding), central or para-central scotomas, peripheral visual field loss with or without arcuate defects, and generalized visual field construction may be present depending on the duration and the severity of the pathology in the retinal nerve fiber layer. Pseudo-papilledema does not typically cause visual field defects as it is not associated with optic nerve deterioration. Therefore, it is very important to differentiate between papilledema and pseudo-papilledema to avoid ultimate complications and waste medical implications by ensuring proper diagnosis and management. Nevertheless, more objective ocular and visual functional diagnostic tests (usually electrophysiological analyses) may be necessary for a more accurate diagnosis in some confusing cases.

- 1. Visual Evoked Potential (VEP): This test measures the electrical response of the brain's visual cortex.
- 2. Pattern Electroretinogram (PERG): This test records the retinal electrical activity to a checkerboard pattern of light and dark squares and provides data about the retinal function.
- **3.** Photopic Negative Response (PhNR) of the Full-Field Electroretinogram (FF-ERG): The test measures the response of the retina to flickering light and gives us important information about retinal dysfunction.

Visual Evoked Potentials (VEPs) are electrophysiological tests used to measure the electrical response of the visual cortex in the brain, which can help detect abnormalities associated with papilledema. The most common VEP abnormality reported in association with papilledema is prolonged p100 latency (transient response) or delayed phase (steady state phase). However, because of this reason, the peak contrast sensitivity may be lower than normal in some papilledema cases.

In pseudo-papilledema, the Visual Evoked Potential (VEP) may be abnormal, reflecting the underlying optic nerve abnormalities. So as a common cause of pseudo-papilledema, optic nerve drusen may cause abnormal VEP. The VEP is an essential test to evaluate the functional integrity and performance of the whole visual pathway. At this point, it can be highlighted here that the delayed or prolonged VEP responses are seen more often within papilledema while the depressed amplitude of undulations may be seen with pseudo-papilledema. The type of VEP testing method mentioned here is the Pattern Reversal VEP (PVEP). The Pattern Onset VEP and Flash VEP testing methods also can be used to detect the nature of the oedematous appearance in these cases. Because of its consistency and reliability, the Pattern Reversal VEP method is the most prominent golden standard technique in the field. Another type of VEP testing is the multifocal VEP (mfVEP) technique which can be used especially for macular area analysis.

In summary, VEP is an important tool for the diagnosis and management of optic nerve disorders such as papilledema and pseudo-papilledema. Abnormal VEPs can provide valuable information about the functional status of the visual pathway and help diagnosis. So, the Visual Evoked Potential may have increased latent period in papilledema patients and decreased amplitude in pseudo-papilledema cases. The recovery of VEP amplitude changes in the resolution of pseudo-papilledema is quite often while the prolonged latent period of VEP may be persistent in papilledema due to irreversible demyelination of the optic nerve.

Management of Clinical Progression

The early and correct diagnosis of papilledema is vital as it may be a forerunner sign of serious underlying conditions such as an intracranial tumor, abscess, or hemorrhage and it may also cause an optic atrophy and total blindness. The prognosis of papilledema depends on the chronicity and severity of the disease. Chronically high intracranial pressure (ICP) can lead to permanent damage to the nerve fiber layer, resulting in progressive visual field defects, deterioration in central visual acuity, and finally can lead to partial or complete vision loss, especially in untreated patients. The treatment for papilledema involves treating the underlying cause of increased intracranial pressure (ICP). If the high pressure of the cerebrospinal fluid is caused by a brain tumor, corticosteroids may be given, but surgery to remove the tumor or radiation therapy to decrease its size may be needed. If papilledema is caused by Idiopathic Intracranial Hypertension (IIH), medications such as carbonic anhydrase inhibitors are strongly recommended, struggle to control the excessive weight may be useful in a long-term strategy. Because the early diagnosis and treatment of papilledema can lead to better outcomes, regular eye examinations are very important to reduce the risk of developing papilledema, especially in patients with predisposing factors. The importance of the correct distinction between true papilledema and pseudopapilledema must be kept in mind in all cases. As a hint in the fundoscopic examination, swelling of the peripapillary nerve fiber layer causes an obscured view of underlying retinal vessels in true papilledema, while the vascular plexus can be seen more clearly in pseudo-papilledema. Additionally, ancillary testing such as B-scan ophthalmic ultrasonography and intravenous fluorescein angiography can be used to differentiate between the two conditions. These tests can also help identify underlying causes of pseudo-papilledema, such as Optic Disc Drusen, myelinated nerve fibers, and high hypermetropia. In some cases, a family history of Optic Disc Drusen may also support the diagnosis of pseudopapilledema. Optic Disc Drusen is mostly asymptomatic and most people with drusen do not know their condition and do not have any symptoms. However, some patients with Optic Disc Drusen may experience some vision problems, including gray or blurry vision for short periods, losing peripheral vision transiently, and seeing white or yellow lumps on the optic discs. Pseudopapilledema is the anomalous elevation of one or both optic discs without edema of the retinal nerve fiber layer, while papilledema is the

swelling of the optic disc due to increased intracranial pressure. Pseudo-papilledema can be caused by various factors such as optic nerve head drusen, congenital anomalies, myelinated nerve fibers, peripapillary masses, and systemic conditions. It is crucial to differentiate between the two, as papilledema can be a sign of a serious medical condition, whereas pseudo-papilledema does not involve increased intracranial pressure and its associated risks. Distinguishing between the two conditions is important to avoid unnecessary invasive testing or surgical interventions. The differential diagnosis between these two matters can be achieved through an exact ophthalmic examination including careful fundoscopic examination with the help of fundus fluorescein angiography, and optical coherence tomography analysis of the retinal nerve fiber layer.

Pseudo-papilledema is defined as an anomalous elevation of one or both optic discs without edema of the retinal nerve fiber layer. The prognosis of Optic Disc Drusen (ODD) in pseudopapilledema is very good. Individuals with drusen develop visual field defects with age, which are usually mild, peripheral, and very slowly progressive. No treatment is necessary for pseudopapilledema if it is related to a congenital variant. If the pseudo-papilledema appearance is due to a peripapillary tumor, the direction of treatment should be at the tumor as appropriate. Patients with optic disc anomalies are recommended to be evaluated frequently by an ophthalmologist, who will monitor for changes such as hemorrhages of the disc or enlargement of scotomas. If the disc anomalies are unchanging and pseudo-papilledema is established, the patient can be informed about that to avoid unnecessary invasive testing or surgical interventions.

The symptoms of pseudo-papilledema may be more significant in some patients including headache, transient vision loss, back pain, pulsatile tinnitus, dizziness, and a distinct pattern of visual field defects. Patients with true papilledema, on the other hand, often present with progressive headaches, nausea, vomiting, and other neurologic symptoms such as positional headaches, transient visual obscurations, and binocular diplopia. It's important to differentiate between pseudo-papilledema and true papilledema, as the causes and implications are different. Pseudo-papilledema is usually benign, and most patients lack visual symptoms, while true papilledema, which is swelling of the optic disc due to increased intracranial pressure, can be very serious and must be treated. A careful history of symptoms of raised intracranial pressure or other neurological symptoms is mandatory to distinguish between the two conditions.

There are no recognized treatment options for pseudo-papilledema caused by Optic Disc Drusen (ODD), which is the most common cause of pseudo-papilledema. Most cases of pseudopapilledema represent normal physiologic variants, and no treatment is needed. However, diseases possibly associated with disc drusen may need treatment, such as subretinal neovascular membrane, central retinal vein occlusion, or ischemic optic neuropathy. If the disc anomalies are unchanging and pseudo-papilledema is established, the patient has to be informed about that to avoid future medical

confusion and complications.

For papilledema due to Idiopathic Intracranial Hypertension, a healthcare provider may prescribe a carbonic anhydrase inhibitor such as acetazolamide. However, this does not apply to pseudopapilledema, which is not due to increased intracranial pressure. Surgery is not a standard treatment for pseudo-papilledema, particularly when it is caused by Optic Disc Drusen (ODD), which is the most common cause. Most cases of pseudo-papilledema represent normal physiologic variants and do not require treatment. However, diseases that can cause pseudopapilledema such as subretinal neovascular membrane, central retinal vein occlusion, or ischemic optic neuropathy must be treated individually. Patients with optic disc anomalies are recommended to be evaluated frequently by an ophthalmologist, who will monitor for changes. It will be very useful to bring back to here that the patients who have no change in the appearance or condition of their pseudopapilledema must be informed about that to avoid any unnecessary invasive tests or surgical interventions. Some surgical procedures may be performed in some cases related to these conditions. For papilledema due to Idiopathic Intracranial Hypertension, surgical procedures may be suggested if other methods are ineffective in relieving and decreasing intracranial hypertension. However, this does not apply to pseudo-papilledema, which is not due to increased intracranial pressure. Radial Optic Neurotomy (RON) is a surgical technique primarily designed to release pressure from the occluded central retinal vein, and it has been proposed for treating central retinal vein occlusion (CRVO) and glaucoma.

Pseudo-papilledema does not require RON intervention, as it often represents a normal physiologic variant. It is fair enough to screen the pseudo-papilledema properly till the progression of the condition. Another surgical procedure is called optic nerve sheath decompression or optic nerve sheath fenestration (ONSF), which can be used to relieve worsening ocular symptoms in cases of medically uncontrolled Idiopathic Intracranial Hypertension, which can cause papilledema. ONSF is the ultimate surgical option to relieve the symptoms of patients who are not responsive to even extensive medical treatment. ONSF procedures can diminish the papilledema and improve the visual symptoms, but it is not a good solution for chronic headaches.

In cases of such persistent headaches, cerebrospinal fluid (CSF) shunting may be helpful. As it is seen, the decision of surgical treatment and type of surgery must be made on a case basis, weighed against the other alternative medical options and implications. The surgical procedures used in papilledema treatment depend on the underlying causes of the condition. Various surgeries including shunt surgeries, craniotomies, and optic nerve sheath fenestration can be done depending on the cause. Shunting surgeries involve the placement of a ventricular catheter into the cerebral ventricles to bypass the flow obstruction. Generally, these shunts drain the fluid into the peritoneal cavity (ventriculo-peritoneal shunt), but alternative sites include the right atrium (ventriculo-atrial shunt), pleural cavity (ventriculo-pleural shunt), etc. Another surgical procedure is the craniotomy which involves the drilling of holes

in the skull to reduce the pressure on the brain by removal of the intracranial hematoma. Pseudo-papilledema is the swelling of the optic nerve head due to various causes. The treatment of this clinical situation depends on the underlying cause. For example, some increased autoimmune inflammatory conditions can be treated with corticosteroids or other immunosuppressive drugs, while infectious causes should be treated with appropriate antibiotics. Antihypertensive medications can be administered in the presence of prominent hypertension. Treating the underlying condition will usually decrease the elevation in the optic nerve head and prevent permanent optic nerve damage. If left untreated, papilledema can lead to permanent and irreversible blindness. In the confusing optic nerve swelling cases, some magnetic resonance imaging (MRI) tests may be necessary to evaluate the intracranial conditions. It should not be forgotten that some certain intra-ocular conditions may cause an oedematous appearance in the optic disc such as retinal vascular occlusions and vitreoretinal tractions. These oedematous changes may also be reversible in the treatment of the underlying ocular condition [1-17].

Conclusion

"Optic nerve" is the name of the visual nerve that takes the longest path within the skull, formed by the axonal extensions of specialized nerve cells of the retina. Raised edematous swelling in which the borders of the optic nerve head can be erased is called papilledema. Although the cause of papilledema cannot often be determined exactly, the real cause can be revealed with a detailed and careful examination. Unlike true papilledema, there is no increase in intracranial pressure in pseudo-papilledema, and therefore, diagnosis and detection are more difficult. It may be very difficult to detect the correct type of oedematous swelling in the optic nerve head utilizing only the routine ophthalmoscopy. However, a more detailed examination, clinical tests, and detailed anamnesis are often required to correct diagnosis. While there are several neurological and/or ophthalmic complaints and findings in true papilledema, there may be any of them in pseudopapilledema. Consequently, distinguishing correctly between these two conditions is very critical for successful treatment and sequel-free outcomes.

References

- 1. John J Chen, M Tariq Bhatti. Papilledema. Int Ophthalmol Clin. 2019; 59: 3-22.
- 2. Gema Rebolleda, Aki Kawasaki, Victoria de Juan, et al. Optical coherence tomography to differentiate papilledema from pseudo-papilledema. Curr Neurol Neurosci Rep. 2017; 17: 1-13.

- 3. Jim Shenchu Xie, Laura Donaldson, Edward Margolin. Papilledema: A review of etiology, pathophysiology, diagnosis, and management. Surv Ophthalmol. 2022; 67: 1135-1159.
- 4. Shikha Talwar Bassi, Kuppuswamy Parthasarthy Mohana. Optical coherence tomography in papilledema and pseudopapilledema with and without optic nerve head drusen. Indian J Ophthalmol. 2014; 62: 1146-1151.
- Angela Guarnizo, Danah Albreiki, Juan Pablo Cruz, et al. Papilledema: A Review of the Pathophysiology, Imaging Findings, and Mimics. Can Assoc Radiol J. 2022; 73: 557-567.
- 6. Madhura A. Tamhankar. The papilledema dilemma. Ophthalmology. 2015.
- Melissa Ko W. Optic Disc Swelling: Papilledema and Other Causes. Neuro-Ophthalmology. 2019; 197-235.
- 8. Jason Zhang, Rod Foroozan. Optic disc edema from papilledema. Int Ophthalmol Clin. 2014; 54: 13-26.
- 9. Ali KUCUKODUK, Sansal GEDIK. Systematic Approch and Ancillary Testing for Diagnosis of Papilledema and Differential Diagnosis from Pseudopapilledema. Ret-Vit. 2021; 30: 104-114.
- Louis Reier, James B Fowler, Mohammad Arshad, et al. Optic Disc Edema and Elevated Intracranial Pressure (ICP): A Comprehensive Review of Papilledema. Cureus. 2022; 14: e24915.
- Amanda Henderson, Andrew Carey R. Controversies in Neuro-Ophthalmic Management. Neuro-Ophthalmic. 2021; 51-63.
- 12. F Grehn, S Knorr-Held, G Kommerell. Glaucomatouslike visual field defects in chronic papilledema. Albrecht Von Graefes Arch Klin Exp Ophthalmol. 1981; 217: 99-109.
- 13. Lin Liu, Michael D Yu, Carol L Shields. Papilledema or pseudopapilledema. Indian J Ophthalmol. 2019; 67: 449.
- Maryam Yadgari, Parivash Nourizade, Nina Firoozian, et al. Crowded Disc in a High Hyperopic Patient Misdiagnosed as Papilledema. Journal of Ophthalmic and Optometric Sciences. 2019; 3: 58-60.
- 15. Paul Freund, Edward Margolin. Pseudopapilledema. StatPearls [Internet]. 2023.
- 16. Olivia M. Crum, Khin P. Kilgore, Rishi Sharma, et al. Etiology of Papilledema in Patients in the Eye Clinic Setting. JAMA Netw Open. 2020; 3: e206625.
- 17. Rishi Sekhri, Helen Kuht, Ian DeSilva, et al. Identifying Biomarkers for Papilledema and Pseudopapilledema. 2023; 64: 23-27.

© 2024 Cuneyt Karaarslan. This article is distributed under the terms of the Creative Commons Attribution 4.0 International License