

Acute Unilateral Vision Loss and Optic Perineuritis (OPN) in Geriatric Patients, a Series of Three Cases

Osama Osman^{1,*}, Julie Gammack¹, Lina Toledo-Franco¹, Takaaki Ishiyama², Emily Reisenbichler³, Shamseldeen Mahmoud⁴, Nongnooch Poowanawittayakom⁵, Connor Clarkston⁶, Cinthia Pi⁷, Andrew J Polk⁷, Osasu N Adah⁷, Jillian Drogin⁸, Sydney O'Neal⁸, Sophie J Xie⁸

¹Department of Internal Medicine, Geriatrics, Saint Louis University, Missouri, USA

²Department of Hospital Medicine, Saint Louis University, Missouri, USA

³Department of Pathology, Saint Louis University, Missouri, USA

⁴Department of Radiology, Saint Louis University, Missouri, USA

⁵Department of Infectious Diseases, Saint Louis University, Missouri, USA

⁶PGY-3 Internal Medicine Resident, Saint Louis University, Missouri, USA

⁷PGY-2 Ophthalmology Resident, Saint Louis University, Missouri, USA

⁸MS-3 Medical Students, Saint Louis University, Missouri, USA.

ABSTRACT

Background: Optic Perineuritis (OPN) is an uncommon condition associated with many systemic disorders. **Methods:** We report three geriatric patients with acute unilateral vision loss and OPN. We also included a brief literature review pertinent to OPN. **Results/Discussion:** While OPN has been reported in association with many conditions such as giant cell arteritis (GCA), it is infrequently associated with mucormycosis sinusitis. Here, we presented cases of OPN associated with both GCA and mucormycosis infection of the sinuses. **Conclusions:** OPN is an uncommon condition that may be idiopathic or secondary to a variety of disorders. Early recognition and urgent treatment of this condition and its underlying causes are necessary to prevent permanent vision loss. Even though many different conditions can lead to OPN, overall, the prognosis of OPN remains good save for mucormycosis associated with OPN where the prognosis is generally poor.

Keywords: Optic perineuritis, Vision loss, Mucormycosis sinusitis, Giant cell arteritis

ABBREVIATIONS: COPD: chronic obstructive pulmonary disease; CRP: C-reactive protein; CT: Computed tomography; DM: diabetes mellitus; ED: Emergency department; ESR: Erythrocyte sedimentation rate; GCA: Giant cell arteritis; IV: intravenous; MRI: magnetic resonance imaging; OD: oculus dexter (right eye); OPN: optic perineuritis; OS: oculus sinister (left eye); RAPD: relative afferent pupillary defect

Vol No: 07, Issue: 02

Received Date: August 25, 2022

Published Date: October 20, 2022

*Corresponding Author

Dr. Osama Osman, MD

Department of Internal Medicine, Geriatrics, Saint Louis University, USA. Phone: 4845157437

E-mail: osama.osman@health.slu.edu

Citation: Osman O, et al. (2022). Acute Unilateral Vision Loss and Optic Perineuritis (OPN) in Geriatric Patients, a Series of Three Cases. Mathews J Ophthalmol. 7(2):28.

Copyright: Osman O, et al. © (2022). This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

BACKGROUND

OPN is an uncommon condition that can be idiopathic or secondary to an underlying cause. In this case series, we report three geriatric patients who suffered from acute unilateral vision loss and optic perineuritis (OPN). The first patient presented with acute unilateral vision loss and OPN of the right eye due to an unknown cause. The second developed OPN in the context of a mucormycosis infection of the sinuses and the third developed OPN secondary to giant cell arteritis (GCA). Most *Mucor* species are commonly found in soil and are unable to infect humans. Thermotolerant species such as *Mucor indicus* sometimes cause opportunistic, and often rapidly spreading, necrotizing infections known as Mucormycosis (previously called zygomycosis). Mucormycosis (zygomycosis) can cause a rare yet aggressive fungal sinusitis in immunosuppressed patients like diabetics, HIV patients, or solid organ transplant patients. Prognosis is generally poor even on the most aggressive intervention, debridement, and intravenous potent antifungal (amphotericin B) [1]. GCA is a systemic medium and large vessel vasculitis that most commonly affects older adults. It is characterized by focal granulomatous inflammation leading to headaches, jaw pain, scalp tenderness, and vision changes. Urgent treatment with high-dose corticosteroids is necessary due to the risk of permanent vision loss. Optic perineuritis can be a rare manifestation of GCA.

METHODS

In this case series, we report three geriatric patients who suffered from acute unilateral vision loss and OPN. The first suffered from acute unilateral vision loss due to OPN of the right eye from an unknown cause. The second developed OPN in the context of inflammatory processes secondary to mucormycosis infection of the sinuses, which was infrequently reported in the past. The third developed OPN from a known autoimmune cause. We also included a brief literature review pertinent to OPN.

Case 1: Idiopathic OPN

A 73-year-old female with a recent history of shingles infection presented to our hospital with right eye vision loss

that began three days before admission. She stated she had decreased vision in the inferior quadrants of her right eye and total loss of her central and superior vision. The patient also endorsed right-sided headache and temporal tenderness without jaw claudication. One week before the presentation, the patient saw her optometrist due to an "odd sensation" in her eye and was prescribed prednisolone eye drops. She saw her retinal specialist on the day of admission, who directed her to the emergency department (ED). Initial labs in the ED were significant for: sodium 132 mmol/L, creatinine 0.36 mg/dL, c-reactive protein (CRP) < 0.5 mg/L, and erythrocyte sedimentation rate (ESR) 20 mm/h. Ophthalmology was consulted for further evaluation.

On the initial ophthalmological exam, visual acuity at near was 20/400 right eye and 20/30 left eye. Intraocular pressures were 12 mmHg in both eyes. An afferent pupillary defect was present in the right eye, and extraocular movements showed a mild limitation in right eye abduction. The external exam was significant for trace inflammatory cells in the vitreous of the right eye, and the dilated fundus exam was significant for mild disc pallor and blurring of the disc margins temporally. An epiretinal membrane was present in the macula of the right eye only, along with paleness to the periphery of her right fundus. Due to concern for GCA, the patient was started on Solumedrol.

MRI of the orbits was obtained, which showed thickening and enhancement of the right optic nerve sheath complex extending to the orbital apex concerning for optic perineuritis (Figure 1). A temporal artery biopsy was obtained, which was negative for GCA. The patient completed 12 doses of IV steroids and was then started on a prednisone taper. A stroke workup was initiated and CT angiography was negative for any arterial occlusion or stenosis. A transthoracic echocardiogram did not show an etiology for branch retinal artery occlusion. Infectious diseases were consulted out of concern of herpes zoster ophthalmicus given her recent shingles outbreak. While in the hospital, she received IV valaciclovir followed by an oral course after discharge. No official diagnosis was made. However, the rheumatology specialists believed giant cell arteritis was the most likely cause of her presentation.

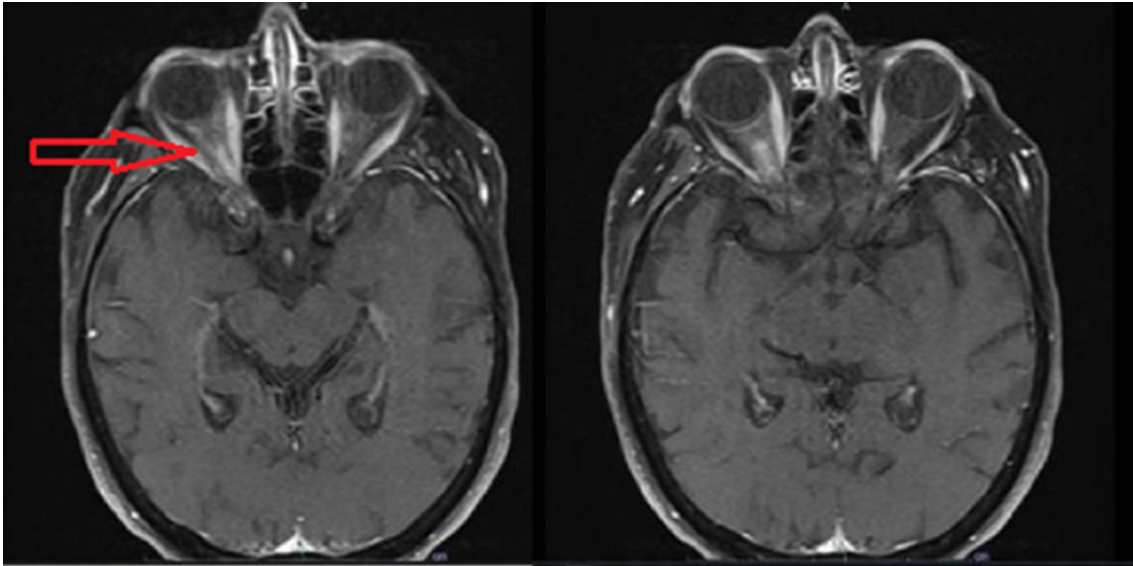


Figure 1: MRI of the Orbits shows thickening and enhancement of the right optic nerve sheath complex extending to the orbital apex, **red arrow**. Differential considerations include optic perineuritis, Tolosa-Hunt syndrome, or optic neuritis/optic neuropathy.

Case 2: Invasive mucormycosis causing OPN

A 75-year-old female with a history of Type II DM and chronic obstructive pulmonary disease (COPD) presented to the ED after a witnessed fall. Her daughter stated that before arrival, her mom had fallen and appeared confused. At baseline, the patient was independent and cared for her medications. However, over the past few weeks, she had a poor appetite and struggled with increased wheezing and shortness of breath due to her asthma. Upon admission, she was diagnosed with diabetic ketoacidosis and started on an insulin drip. Further workup demonstrated leukocytosis and proptosis of her right eye.

On ophthalmologic evaluation, visual acuity at near was 20/70 in the right eye (OD) and 20/40 in the left eye (OS). Intraocular pressures were 29 mmHg OD and 19 mmHg OS. Pupils were unequal and minimally reactive to light. Extraocular movements OD were limited (-3) in all directions of gaze, and OS were limited (-1) in elevation only. The external exam was significant for the prominence of the right globe, complete right ptosis at rest, and mild conjunctival chemosis. Dilated fundus exam revealed normal optic nerves bilaterally, with flushing of the vessels at the

optic disc with slight globe pressure. The cup-to-disc ratio was 0.5 bilaterally, and the remainder of the fundus exam was otherwise unremarkable.

Contrast-enhanced MRI of the orbits showed diffuse paranasal sinus disease in a pattern suggestive of acute sinusitis concerning fungal infection and right OPN. Dehiscence of the lateral nasal wall and medial orbital wall with abscess formation was additionally noted (Figure 2A). ENT was consulted for surgical evaluation, and they performed a bilateral functional endoscopic sinus surgery with cultures showing MRSA, *Klebsiella pneumoniae*, and *Enterobacter cloacae*. Pathology revealed an invasive fungal infection, and the patient was started on IV Amphotericin B (Figure 2B). The patient did not wish to proceed with future debridements after the initial surgery and instead elected to continue with antifungal administration and repeat imaging every two weeks. She reported a loss of vision in her right eye three days after surgery. A repeat MRI revealed post-surgical swelling compressing her optic nerve. Ophthalmology and ENT deferred from starting steroids at this time due to extensive infection. Due to poor prognosis and patient and daughter preference, the patient was transitioned to comfort measures and was discharged home on hospice.

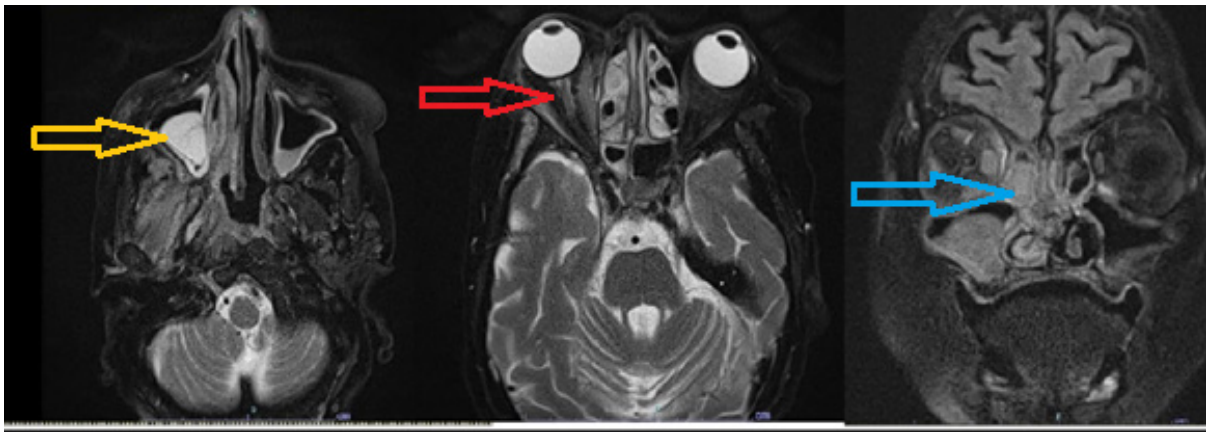


Figure 2A: MRI of the orbits demonstrated invasive fungal sinus infection, **yellow arrow**. Right-sided optic perineuritis was noted red arrow and dehiscence of the lateral nasal and medial orbital wall. A 1 x 0.4 x 1.4 cm abscess in the right orbit extraconal and medial to the medial rectus muscle was also seen **blue arrow**. Mild intraorbital fat infiltration, diffuse hyperenhancement and enlargement of the right-sided extraocular muscles, and proptosis were present.

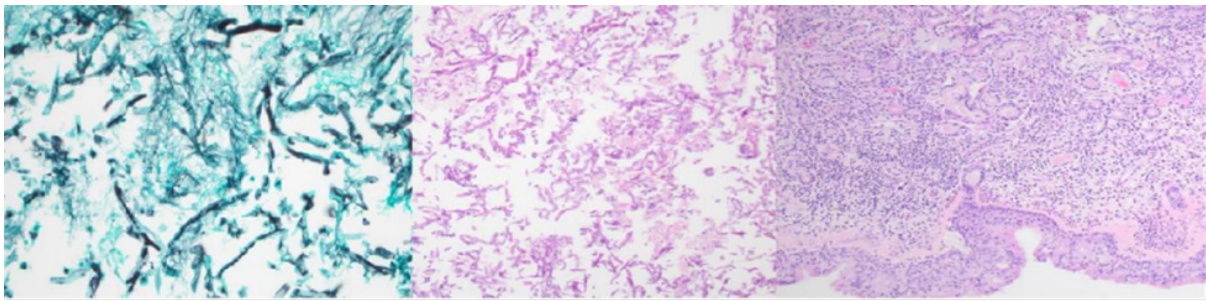


Figure 2B: H&E and GMS sections demonstrate marked submucosal acute and chronic inflammation and fungal infection as evidenced by broad hyphae with variable angle branching.

Case 3: Giant Cell Arteritis causing OPN

A 74-year-old female with no significant past medical history presented to the ED with a six-week history of right eye pain, headaches, and worsening vision. She had previously seen a local neurologist who diagnosed trigeminal neuralgia, for which she was started on prednisone and carbamazepine. However, despite these medications, her vision progressively worsened, and she began to experience diplopia. Within a week of the presentation, she could only perceive light and completely lost color vision. Her neurologist ordered an MRI of the orbits, which showed right exophthalmos, diffuse infiltrative edema, and contrast enhancement throughout the intraconal right orbit with extensive perineural contrast enhancement of the retrobulbar right optic nerve and enlargement of the superior right rectus muscle. In light of the MRI results, she was referred to the ED for a complete workup.

At the ED, an initial ophthalmologic evaluation showed visual acuity of light perception in the right eye (OD) and 20/30 left

eye (OS). Intraocular pressures were 27 mmHg OD and 14 mmHg OS. A pupillary exam was significant for a right eye relative afferent pupillary defect. Extraocular movements revealed complete limitation in all directions of gaze in the right eye and full extraocular motilities in the left eye. An external exam was significant for right proptosis, inability to open the right eye without assistance, and trace chemosis and injection of the right conjunctiva. A dilated eye exam was significant for trace temporal pallor of the right optic disc with an optic cup-to-disc ratio of 0.2 OD and 0.25 OS. The fundus exam was otherwise unremarkable.

On admission, she was hemodynamically stable. Initial labs were notable for CRP 3.7 mg/L and ESR 105 mm/h. She described her right eye pain as stabbing and constant and could only see light from her right eye. Repeat MRI of the orbits was consistent with OPN (Figure 3). The patient was started on high-dose IV methylprednisolone with a slow improvement in her vision and eye pain. A temporal artery biopsy was obtained a week after admission that showed

~50% loss of internal elastic membrane and no active inflammation compatible with healing or treated giant cell arteritis. She was eventually discharged home on a steroid

taper. Two weeks post-discharge, her vision continued to improve and her eye pain resolved.

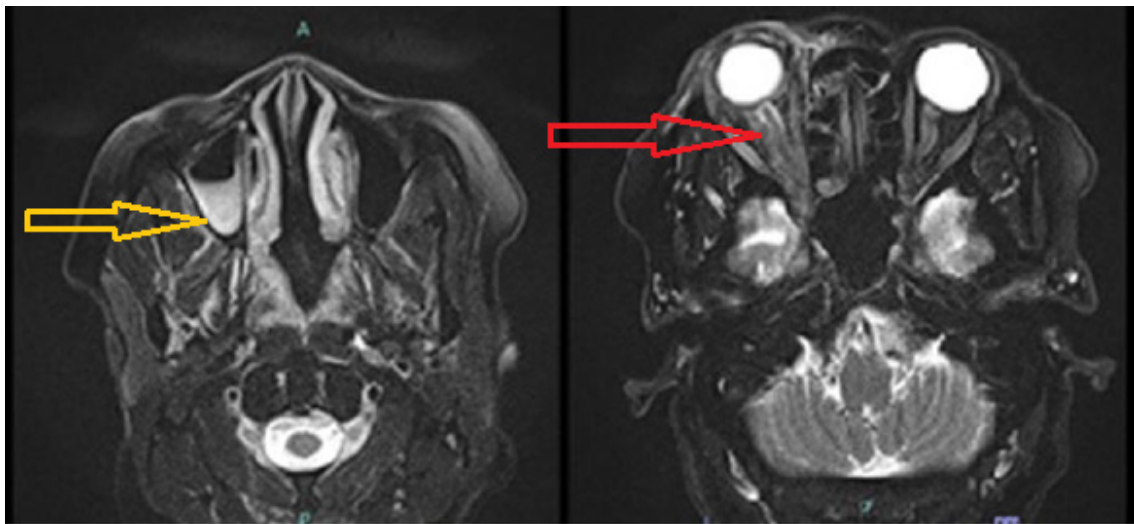


Figure 3: MRI of the orbits showed significant thickening and enhancement of the right optic nerve sheath with surrounding retrobulbar fat stranding extending to the orbital apex, **red arrow**, mild prominence, enhancement of the extraocular muscles, and mild right eye proptosis. Right-sided maxillary sinus infection was noted **yellow arrow**.

RESULTS/DISCUSSION

Optic perineuritis (OPN) is an uncommon inflammatory disorder in which the inflammation is confined to the optic nerve sheath. While most cases are idiopathic, OPN is associated with many systemic disorders including sarcoidosis [2-4], IgG4-related disease [5], granulomatosis with polyangiitis [3,6], giant-cell arteritis [7], Behcet's disease [8], systemic lupus erythematosus [1], inflammatory bowel disease [9], syphilis [3], tuberculosis [3], herpes simplex virus [10], herpes zoster virus [11], leukemia [12], other viral encephalitides [13], and primary or metastatic malignancies [14]. Furthermore, it has also been documented to present simultaneously with neuroretinitis [15]. To our knowledge, very few cases have reported OPN as a complication of mucormycosis sinusitis, as discussed in our case. Mucormycosis is a rare, aggressive, invasive fungal infection that usually afflicts immunosuppressed patients. Prognosis is generally poor even with the most aggressive interventions, including debridement and IV Amphotericin B. Thus, early intervention is crucial [1].

Pathophysiology

The pathophysiology behind OPN may reflect the underlying causes of the condition. Immune-mediated inflammation is primarily confined within the nerve sheath. However,

varying degrees of optic nerve axonal involvement may still occur. This variable degree and location of inflammation contribute to the range of symptoms and exam findings.

Signs/Symptoms

The clinical presentation of OPN may mimic optic neuritis. Most patients present with acute visual loss, eye pain, or both. Like optic neuritis, the eye pain may be exacerbated by eye movement, but the pain in OPN may be more severe or last longer than the typical optic neuritis. The visual loss in OPN is variable and may range from none to severe. Patients may describe visual blurring, dimming, "spots" in vision, or splotches [15]. A phenomenon known as "amaurosis fugax," or transient loss of vision, is also frequently associated with OPN.

Patients with OPN may present with normal-appearing optic discs, optic disc edema, or peripapillary atrophy [Bergman]. Visual field defects can be variable - arcuate defects, paracentral or central scotomas, peripheral island, and altitudinal defects have all been described. Given involvement of the optic nerve sheath and orbit, OPN can also cause orbital signs such as ptosis, ophthalmoplegia, and exophthalmos [3]. While rare, retinal necrosis, scleritis, and episcleritis have been reported as presenting findings in OPN as well [16]. In patients who are non-responsive

to corticosteroids, cases have been reported of the need to perform optic nerve biopsy to confirm the etiology of OPN [17]. Biopsy of OPN will typically show inflammatory infiltration into the dural sheath or perineural fibrosis.

Clinical diagnosis

OPN is often clinically difficult to distinguish from optic neuritis and, in some cases, from other orbital inflammatory or infectious conditions due to the overlap in symptomatology. However, it is essential to identify underlying treatable causes. Clinical similarities to optic neuritis include decreased visual acuity, the possible presence of a relative afferent pupillary defect (RAPD), visual field loss, and dyschromatopsia. However, there are often clinical differences between OPN and optic neuritis. These include faster onset in optic neuritis (days) compared to OPN (weeks). Central vision is usually spared in OPN, leading to milder dyschromatopsia and sporadic or more subtle RAPD. The diagnosis of OPN is typically based on a combination of clinical and radiographic findings. Gadolinium-enhanced, fat-saturated T1 MRI of the orbits is key to diagnosing OPN [15].

General treatment and Prognosis

Initial treatment for OPN generally includes high-dose systemic corticosteroids. Typically, the higher the frequency of flare-ups, the higher dosage of corticosteroids required [15]. In case studies comparing various dosing of corticosteroids, those that used 30-40 mg/day compared to 60-80 mg/day showed higher rates of recurrence. A long and slow taper may be required to decrease the risk of rebound attacks. While there are rare cases of resolution without steroid treatment, OPN typically has a good prognosis with treatment. Unlike demyelinating optic neuritis, there is no known association between OPN and multiple sclerosis. Visual outcomes are also excellent in patients with OPN, with most patients in clinical case studies returning to 20/20 vision or better [15].

CONCLUSIONS

OPN is an uncommon condition with a relatively good prognosis that may be idiopathic or secondary to an underlying cause. Invasive infections involving the sinuses, as seen in Case 2, have infrequently been previously recognized as causes of OPN. Mucormycosis manifests in different ways in immunocompromised patients and those with diabetes mellitus. Devastating rhino-orbital-cerebral and pulmonary infections are the most common syndromes caused by these fungi. Prognosis is generally poor, even with aggressive interventions, including debridement and IV amphotericin

B. Multimodal therapy with surgical debridement and antifungal chemotherapy is required for an optimal outcome. Additionally, if possible, discontinuing immunosuppressive therapy is the cornerstone of management.

REFERENCES

1. Odessey E, Cohn A, Beaman K, Schechter L. (2008). Invasive mucormycosis of the maxillary sinus: extensive destruction with an indolent presentation. *Surg Infect (Larchmt)*. 9(1): 91-98.
2. Purvin V, Kawasaki A, Jacobson DM. (2001). Optic Perineuritis: Clinical and Radiographic Features. *Arch Ophthalmol*. 119(9): 1299-1306.
3. Bergman O, Andersson T, Zetterberg M. (2017). Optic perineuritis: a retrospective case series. *International medical case reports journal*. 10: 181-188.
4. Yu-Wai-Man P, Crompton DE, Graham JY, Black FM, Dayan MR. (2007). Optic perineuritis as a rare initial presentation of sarcoidosis. *Clin Exp Ophthalmol*. 35(7): 682-684.
5. Hickman SJ. (2016). Optic Perineuritis. *Curr Neurol Neurosci Rep*. 16(2): 16.
6. Takazawa T, Ikeda K, Nagaoka T, et al. (2014). Wegener granulomatosis-associated optic perineuritis. *Orbit (Amsterdam, Netherlands)*. 33(1): 13-16.
7. Morotti A, Liberini P, Padovani A. (2013). Bilateral optic perineuritis as the presenting feature of giant cell arteritis. *BMJ case reports*. 2013.
8. Lai C, Sun Y, Wang J, et al. (2015). Optic Perineuritis in Behcet Disease. *J Neuroophthalmol*. 35(4): 342-347.
9. McClelland C, Zaveri M, Walsh R, Fleisher J, Galetta S. (2012). Optic perineuritis as the presenting feature of Crohn disease. *J Neuroophthalmol*. 32(4): 345-347.
10. Byon IS, Jung JH, Kim HY, Park SW, Lee JE. (2013). Optic perineuritis secondary to acute retinal necrosis. *J Neuroophthalmol*. 33(4): 419-421.
11. Townsend JH, Dubovy SR, Pasol J, Lam BL. (2013). Transient optic perineuritis as the initial presentation of central nervous system involvement by pre-B cell lymphocytic leukemia. *J Neuroophthalmol*. 33(2): 162-164.
12. Miller N, Newman N, Biousse V, Kerrison J. (2005). Walsh & Hoyt's Clinical Neuro240 Ophthalmology, 6th Edition. Philadelphia, PA: Lippincott Williams & Wilkins. 241: 337-338.

13. Hickman SJ. (2016). Optic Perineuritis. *Curr Neurol Neurosci Rep.* 16(2): 16.
14. Wals KT, Ansari H, Kiss S, Langton K, Silver AJ, Odel JG. (2003). Simultaneous occurrence of neuroretinitis and optic perineuritis in a single eye. *J Neuroophthalmol.* 23(1): 24-27.
15. Purvin V, Kawasaki A, Jacobson DM. (2001). Optic Perineuritis: Clinical and Radiographic 247 Features. *Arch Ophthalmol.* 119(9): 1299-1306.
16. Zaidi S. (2013). Optic Perineuritis: Uncommon presentation of more common diseases. *Journal of Neurology, Neurosurgery & Psychiatry.* 84(11): e2
17. Dutton JJ Anderson RL. (1985). Idiopathic inflammatory perioptic neuritis simulating optic nerve sheath meningioma. *Am J Ophthalmol.* 100(3): 424-430.