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CASE REPORT

Compressive Optic Neuropathy (CON) Secondary to Inflammatory Polyp Mimicking Lymphoma: A Rare Manifestation

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Abstract

Introduction: Compressive optic neuropathy (CON) is the impairment of optic nerve function by space-occupying lesions that mechanically compress the optic nerve or optic tract. CON can arise from either intrinsic or extrinsic compression, and determining the underlying cause is important to approach the correct management. The objective of this case report is to present a rare case of CON secondary to an inflammatory polyp that mimicking lymphoma. **Case Presentation:** A 68-year-old male patient came to the neuro-ophthalmology clinic with a chief complaint of blurry vision in his left eye since three months ago that was getting worse until he lost his vision accompanied by drooping of the upper left eyelid. He also felt pain in his left eye, nausea, vomiting, and headache. Visual acuity of the right eye was 5/5 and the left eye had no light perception. Ocular motility of the left eye was restricted in all gaze. The pupil was 6 mm with no light reflex, and edema in the optic nerve head in the left eye was found from the posterior segment. The patient's MRI showed soft tissue thickening on the left orbital apex – left optic canal – left anterior cavernous sinus and enlargement of left medial rectus muscle with encasement of the left optic nerve, and from post-contrast showed contrast enhancement that suggested a lymphoma. He was diagnosed with CON secondary to lymphoma and was given Methylprednisolone 32 mg three times a day orally. **Conclusions:** Several differential diagnoses resemble the findings of CON secondary to inflammatory polyp. Some diagnostic examination must be done to exclude the other diagnosis to give a proper treatment for the patient.

Keywords: compressive optic neuropathy (CON); lymphoma; inflammatory polyp

Introduction

Compressive optic neuropathy (CON) refers to the functional deterioration of the optic nerve due to space-occupying lesions that exert mechanical pressure on either the optic nerve or the optic tract. The documented incidence of CON, based on reported cases, stands at approximately four cases per 100.000 individuals annually. The distribution of CON concerning gender, race, ethnicity, and age is contingent upon the particular underlying cause or etiology.[1] The occurrence of CON can arise from either intrinsic or extrinsic compression spanning any segment of the optic nerve. Beyond the impact induced by compression, optic nerve impairment can manifest due to factors including demyelination, ischemia, metabolic dysfunction, and traumatic injury. Among the prevailing clinical indicators, a gradual, progressive loss of vision in one eye is a prevalent feature, occasionally accompanied by headache symptoms. Bilateral visual decline can emanate from midline lesions like pituitary adenomas, craniopharyngiomas, meningiomas, large aneurysms, or bilateral orbital lesions such as thyroid disorders or sarcoidosis. Accurate determination of the underlying cause of CON is paramount, as the diagnostic landscape encompasses a wide spectrum, necessitating varied management approaches.[2],[3]

Nasal polyps are believed to emerge due to inflammation of the nasal mucosa, often associated with allergies or anatomical obstructions caused by altered air

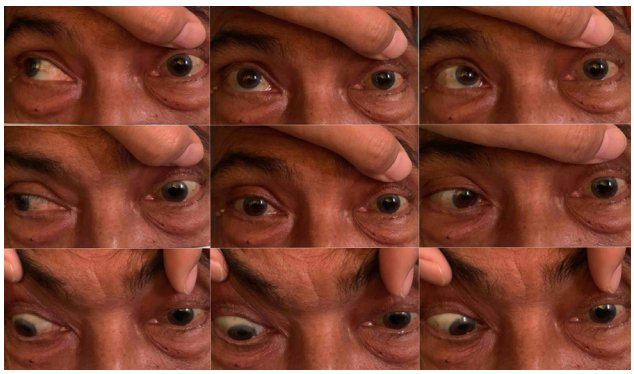


Figure 1. Patient's nine gaze showed limitation of movement in the left eye.

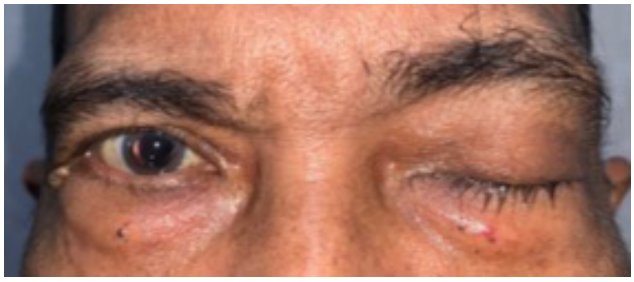


Figure 2. Patient's left eye showed severe ptosis with IPF, MRD1, MRD2, MLD and LF were 0 mm.

currents and increased air pressure. These polyps can originate from the mucosal lining of both the nose and the paranasal sinuses. The sinuses most frequently affected by these polyps are the ethmoid sinuses, followed by the maxillary, sphenoid, and frontal sinuses, in that sequential order. Given the anatomical proximity of the optic nerve to the paranasal sinuses, several factors, including inflammation and hypoxic conditions within the sinuses, could potentially exert structural and functional influences on the adjacent optic nerve. However, several differential diagnoses can be the underlying cause of CON with similar signs and symptoms. In this case report, we will present a 68-year-old male who was diagnosed with suspected CON secondary to an inflammatory polyp that was mimicking lymphoma.[4],[5]

Case presentation

A 68-year-old male was referred to the neuro-ophthalmology unit at Dr. Soetomo General Hospital, Surabaya, Indonesia, with a chief complaint of blurry vision in the left eye since four months ago. Blurry's vision was worsening slowly until his vision became dark two months ago. The patient has a history of cataract surgery in his right eye five months ago and left eye a month later, furthermore, he said his left eye vision was never clearer than before the surgery. After three weeks of surgery, the complaint developed. The complaint was accompanied by pain in the left eye. The patient also complained of nausea, vomiting, headache, and nasal congestion. There was no history of eye redness and decreased visual field. The patient has no history of trauma, allergy, hypertension, diabetes mellitus, or similar complaints in his family.

Physical examination revealed that he was compos mentis. His blood pressure was 120/80 mmHg, heart rate 88 bpm, respiratory rate 20 bpm, and body temperature was 36.5°C. Motoric examinations in the right-left arm and right-left leg were found five in all extremities, and sensory examinations found no hypoesthesia. Cranial nerve examinations showed no signs of lateralization. An ophthalmology examination revealed his visual acuity was 6/6 in the right eye and no light perception in the left eye. The intraocular pressure in both eyes was 10 mmHg.

Color vision examination using the Ishihara test was 38/38 plate on the right eye and the left eye could not be evaluated. His right eye movement was within normal limits, while his left eye had limited movement -4 to superolateral, lateral, inferolateral, and -3 to superior, superomedial, medial, inferomedial, and inferior, with pain during the movement (Figure 1). The worth 4-dot test could not be evaluated.

From ptosis examination, we found in right eye interpalpebral fissure (IPF) was 9 mm, MRD1 (margin reflex distance) was 4 mm, MRD2 was 5 mm, margin limbal distance (MLD) was 9 mm, levator function (LF) was 12 mm. Left eye was found severe ptosis with IPF, MRD1, MRD2, MLD and LF were 0 mm (Figure 2). Slitlamp biomicroscopy examination of the right eye's anterior segment revealed a pupil with a 3 mm diameter, normal light reflex, and pseudophakic. We found a pupil diameter of 6 mm from the left eye with no light reflex and pseudophakic (Figure 3).

Posterior segment examination on the right eye found positive fundal reflex, optic nerve head with defined margin and slight hyperemia color, cup disc ratio 0.2. From the left eye was found positive fundal reflex, optic nerve head with blurry margin in all quadrants, and hyperemia with the cup disc ratio 0.1 (Figure 4). We performed an OCT examination and found optic disc on both eyes was atrophy, the macula showed slight thinning in the right eye, and from ganglion cell complex showed thinning in the inferior and inferonasal quadrant of the left eye (Figure 5). We also performed a visual field examination and found a defect in the superior and nasal quadrant of the right eye (Figure 6).

The patient's blood laboratory tests were done to evaluate systemic disease and to consider treatment options. We found leucositosis (white blood cells (WBC)

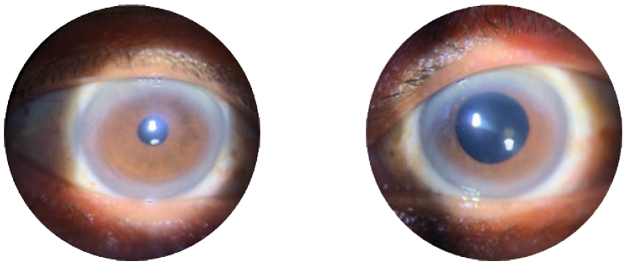
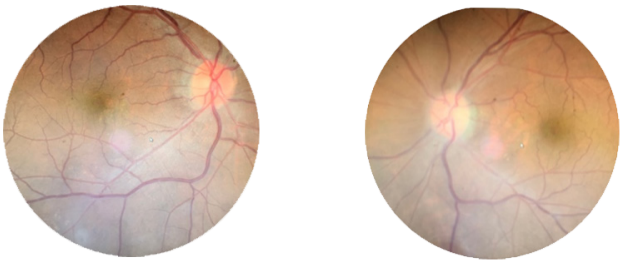


Figure 3. Anterior segment examination showed the pupil in the left eye with a diameter of 6 mm.



A B

Figure 4. Funduscopic photograph (A) Right eye and (B) Left eye.

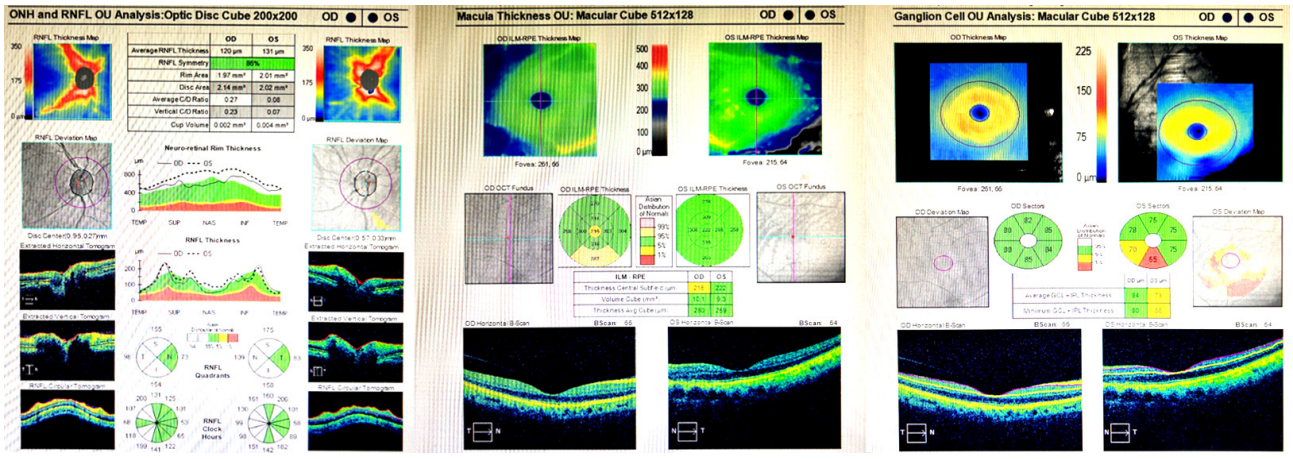
10.56 x 103), elevated low-density lipoprotein (LDL) value (129 mg/dL) and hyponatremia (Na 129 mmol/L). He also had undergone an imaging study, head computed tomography (CT) scan, and magnetic resonance imaging (MRI) brain. A head CT scan was performed from the previous hospital, and pansinusitis was found. From MRI, the brain without contrast was found with soft tissue thickening on the left orbital apex – left optic canal – left anterior cavernous sinus and enlargement of left medial rectus muscle with measurement 1.5 x 1.5 x 3 cm with encasement of the left optic nerve. The post-contrast showed contrast enhancement, hypointense in T2, hyperintense in diffusion-weighted imaging (DWI) and hypointense in apparent diffusion coefficient (ADC), suggesting a lymphoma. We also found minimal microvascular ischemia in the subcortical frontal and left-right parietal. From magnetic resonance angiographic (MRA), fenestrated basilar artery (BA), and hypoplasia left ventricular aneurysm (LVA), that could be an anatomic variant (Figure 7).

For initial planning, we gave methylprednisolone oral 32 mg tab every 12 hours, omeprazole 20 mg tablet every 12 hours, neurotropic one tablet every 12 hours, and folic acid one tablet every 12 hours. After a one-week follow-up, the patient complained that the pain had been significantly reduced, but there was no improvement in visual acuity. We consulted the medical oncology department and planned to undergo abdominal and lymph node ultrasound, and the result was normal, with no enlargement in the lymph node. After that, the patient was consulted by the reconstruction, oculoplasty, and oncologist (ROO) department, and they concluded the diffuse mass in the orbital apex area was suspected as either inflammation sinusitis or lymphoma process; then they diagnosed the patient with CON and left eye painful blind eye. We also consulted the otorhinolaryngology department. The patient was diagnosed with chronic sinusitis, and functional endoscopic sinus surgery (FESS) was performed. Intraoperatively, a mass was found in the ethmoid sinus, and a biopsy incision was made. The histopathology result showed an inflammatory polyp.

Discussion and conclusions

CON denotes the optic nerve's functional impairment resulting from space-occupying lesions that exert mechanical pressure upon either the optic nerve or the optic tract. Depending on their locations, these compressive optic neuropathies can be categorized into anterior and posterior forms. The specific appearance is contingent upon factors such as the temporal alignment, spatial position, and duration of the compression. Notably, the presence of a swollen optic disc signifies that the compression site is situated within or proximate to the orbital apex.[6],[7],[8]

Individuals affected by CON typically manifest a gradual and progressive decline in visual acuity, which can occur in one or both eyes. Presenting symptoms may encompass a range of manifestations, including headaches, nausea, vomiting, diplopia, dyschromatopsia, exophthalmos, afferent pupillary defect, photophobia, redness of the eye, or unexplained weight loss. The state of the optic disc in an eye afflicted by compressive neuropathy may exhibit variations, appearing either normal, edematous (swollen), or atrophic.[1] From this case report, we found the patient with progressive monocular vision loss four months ago with nausea, vomiting, and headache. There was also severe ptosis with limitation



A B C

Figure 5. (A) Optical coherence tomography (OCT) of the optic disc showed atrophy in the optic nerve head in both eyes; (B) Macula OCT showed slight macula thinning in the right eye; and (C) Ganglion cell complex in the left eye showed thinning in the inferior and inferonasal quadrant.

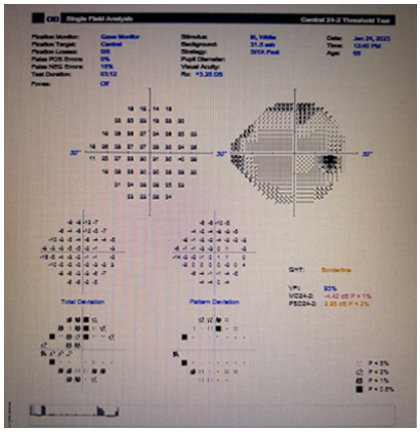
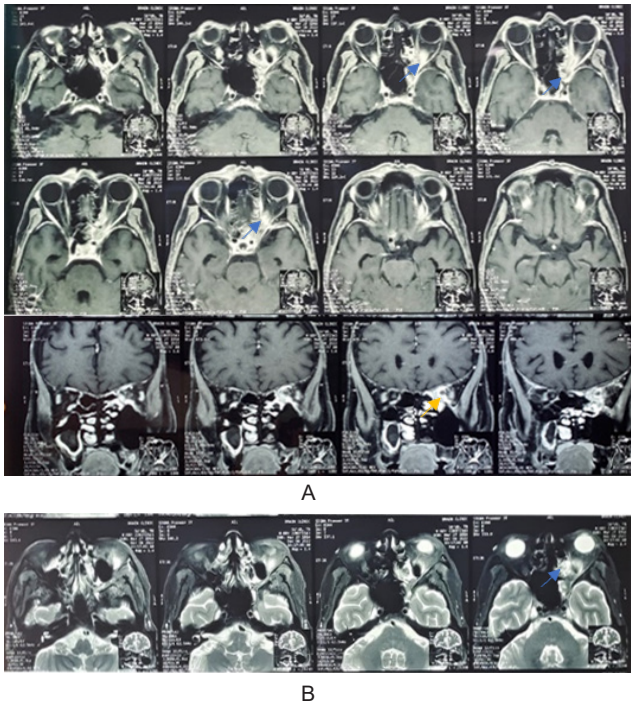


Figure 6. Humphrey's Visual Field (HVF) of the right eye showed visual field defect in superior and nasal.

of ocular motility in all directions. Posterior segment examinations showed optic disc edema in the left eye.

Lymphomas are a heterogeneous group of malignancies that arise from the clonal proliferation of B-cell, T-cell, and natural killer (NK) cell subsets of lymphocytes at different stages of maturation. Approximately 10% of orbital malignant lymphomas show predominant involvement of the extraocular muscle (EOM), which rarely results in CON.[9],[10] The patient was initially diagnosed with CON secondary to lymphoma. This diagnosis was also supported with the patient's MRI brain without contrast, and we found soft tissue thickening on the left orbital apex – left optic canal – left anterior cavernous sinus and enlargement of left medial rectus muscle with measurement 1.5 x 1.5 x 3 cm with encasement of the left optic nerve. The post-contrast showed contrast enhancement, hypointense in T2, hyperintense in DWI, and hypointense in ADC, suggesting a lymphoma (Figure 7). Another report with a similar case found enlargement of the right medial and inferior rectus muscles with their tendons, compressing the optic nerve at the orbital apex from the patient's MRI. T1W (longitudinal relaxation time) and T2W (transverse relaxation time) images showed isointense enlargement of the right medial and inferior rectus muscles concerning grey matter. T1W images after gadolinium (Gd) administration showed enhancement of the margin of both rectus muscles, including adipose tissue around these muscles; however, they showed no enhancement of the optic nerve or its sheath. The T1W-Gd images also demonstrated possible extension into the cavernous sinus through the superior orbital fissure.[11]

A lymphoma diagnosis is confirmed through a tissue biopsy, a procedure that involves obtaining a sample of the affected tissue for analysis. Several techniques are commonly employed for this purpose, including fine-needle aspiration, core biopsy, incision or wedge biopsy, and excisional biopsy. Among these, the excisional biopsy is often regarded as the "gold standard" due to



A

B

Figure 7. MRI brain and orbital focused examination (A) Without contrast was found soft tissue thickening on the left orbital apex – left optic canal – left anterior cavernous sinus and enlargement of left medial rectus muscle (blue arrow) with encasement of the left optic nerve (yellow arrow) and (B) Post-contrast showed contrast enhancement that suggested a lymphoma (yellow arrowhead).

its ability to evaluate the entire lymph node architecture comprehensively.[12] Our patient has undergone biopsy from the posterior ethmoidal sinus, and the histopathology result showed polypoid epithelial stacked with round nuclei, smooth chromatin, cytoplasm, and stroma composed loosely with lymphocyte, plasm cell, neutrophil, and macrophage, suggesting an inflammatory polyp.

Polyps can be developed within the paranasal sinuses, along with the presence of inflammation-induced mucosal thickening that characterizes chronic sinusitis.[13] The factors influencing chronic rhinosinusitis encompass a wide range, spanning genetic predisposition, immune system disorders, impaired mucociliary function, and structural irregularities within the nasal cavity and paranasal sinuses. In addition to these factors, the persistence of a bacterial infection in the sinuses that is inadequately managed can lead to the progression of acute sinusitis into the chronic form. This ongoing inflammation can potentially expand and exert pressure on neighboring structures and create conditions of reduced oxygen availability (hypoxia).[14] In this case, the patient was also diagnosed with pansinusitis from the ENT department. However, the signs and symptoms of the patient were similar to lymphoma, so we diagnosed the patient with CON secondary to lymphoma. Treatment of polyps includes an adequate trial of corticosteroids, and we started to give corticosteroid therapy, but there was no visual improvement. In the case of resistant polyps, endoscopy sinus surgery is the treatment of choice.[4]

In this context, it is imperative to acknowledge a range of differential diagnoses that may exhibit clinical findings similar to the patient's. Consequently, a series of diagnostic assessments must be performed to rule out alternative potential diagnoses effectively. These comprehensive evaluations are essential to accurately determine the appropriate course of treatment for the patient's condition.

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