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HOLISTIC APPROACH IN BILATERAL VISUAL LOSS AND OPHTHALMOPLEGIA CASE DUE TO MULTICRANIAL NERVE PALSY

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ABSTRACT
Background: Ophthalmoplegia refers to the paralysis or weakness of extraocular muscles that have numerous etiologies and different clinical presentations based on the anatomical lesion. In some cases, ophthalmoplegia may also be accompanied by decreased visual acuity. Loss of visual acuity in both eyes is rare, and several workups may be necessary.
Case Presentation: A 34-year-old man came to a tertiary hospital. He complained of vision loss in both eyes. Restricted eye movement and proptosis were found on ophthalmology examination. Based on MRI and CT scan there were found sinonasal mass that compress optic nerve and muscle, but from biopsy was found granulomatous chronic inflammation which lead to tuberculosis.
Conclusion: Bilateral vision loss and ophthalmoplegia rarely happen and the etiology may vary. Imaging and the histopathology exam can guide to the definitive etiology and help the diagnosis, so the patient could get a proper treatment.
Keywords: Compressive optic neuropathy, ophthalmoplegia, sinonasal mass

BACKGROUND

Ophthalmoplegia is defined as paralysis or weakness of one or more of the muscles that control eye movement. Ophthalmoplegia can be caused by injury or disease of the cerebral hemispheres, midbrain, pons, and cerebellum. The cranial nerves responsible for ocular movement can be affected intrinsically or extrinsically throughout their course in the brainstem, skull base, cavernous sinus, and orbit. [1,2]

Ophthalmoplegia is divided into external ophthalmoplegia, internal ophthalmoplegia, and total ophthalmoplegia. External ophthalmoplegia refers to paralysis of the extraocular eye muscles which function to move the eyeball. Internal ophthalmoplegia is a combination of paralysis of the constricting pupils (iridoplegia), which causes mydriasis, and cycloplegia, which causes loss of power (inability to see near), which muscles are innervated by the autonomic nervous system. [1,2]

While the third is total ophthalmoplegia. The term total ophthalmoplegia is the complete loss of function of all eye muscles, both intrinsic and extrinsic; where total weakness is obtained in N.III which is characterized by the presence of ptosis, iridoplegia, cycloplegia, and paralysis of all the extra-ocular muscles innervated by N.III which also involve the lateral and superior oblique muscles.[2]

In some cases, ophthalmoplegia may also be accompanied by decreased visual acuity. Loss of visual acuity in both eyes is rare and can be caused by multifactorial etiologies. In this case report, we will discuss how to establish the diagnosis and differential diagnosis in patients with decreased visual acuity and ophthalmoplegia symptoms.

CASE PRESENTATION

A 34-year-old man came to the Neuro-Ophthalmology Division complaints of both eyes blurred vision in in the last 3 weeks. Right eye ptosis was noticed one week before the vision loss.

For systemic complaints, the patient complained of nasal congestion for the past month, and the patient also had greenish-

yellow mucus from the nose several times during the past month, sometimes accompanied by blood. There was also a significant decrease in smell and the patient felt his voice had become nasal for the past 1 month. The patient denied weakness in the extremities.

On physical examination, the patient's right visual acuity is light perception, while in the left eye, the patient had no light perception vision. On examination of the anterior segment of both eyes, the pupil diameter of the right eye was 4mm with a slow pupil reflex and the pupil diameter of the left eye was 6 mm with no pupil reflex, while the other anterior segments were within normal limits. on examination of the posterior segment using funduscopic within normal limits.

Upon examination, the right eye revealed complete limitation in every direction. The mobility of the left eye was entirely confined, except the medial and inferomedial regions, which were slightly limited. Forced duction test revealed no restriction. A palpebral examination was performed, and a small difference was found between the vertical palpebral fissures of the right eye 7 and the left eye 9, and for other palpebral examination parameters within normal limits. On examination of the cranial nerves, examination of N.II function could not be carried out because of the patient's visual acuity, CN.V and CN.VII were normal. Bilateral proptosis were examined using Hertel Exophthalmometer, we found 22 mm in the right eye and 23 mm in the left eye.

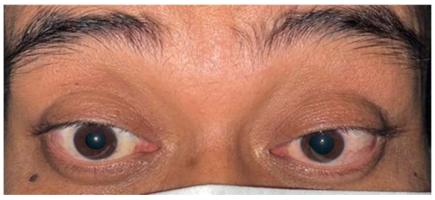

Figure 1. Proptosis on both eyes

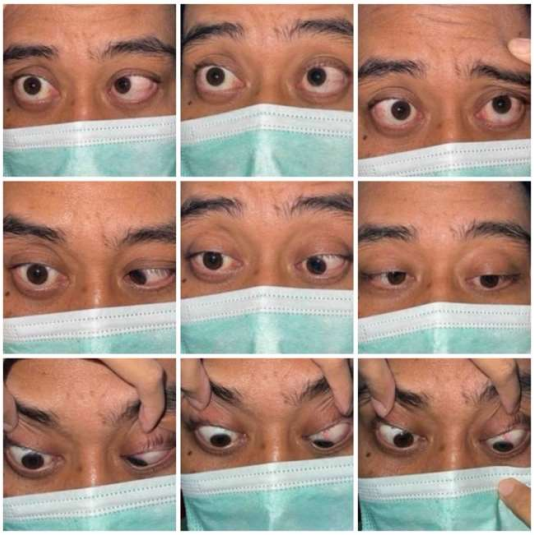

Figure 2. Limitation of eye movements on both eyes

The patient was then diagnosed as having bilateral proptosis and ophthalmoplegia with compressive optic neuropathy. The patient was then planned to perform head and orbit's MRI with and without contrast, as well as complete blood laboratory tests and hormonal test.

All laboratory test results were found to be within normal limits, except for CD4 which was 267 cells/μL with non reactive result of anti-HIV rapid test. The patient was diagnosed with suspected idiopathic lymphocytopenia, and no specific management in the field of internal medicine.

Visual acuity of both eyes had decreased to no light perception. Disc swelling of both eyes were seen on funduscopy examination. OCT of the optic nerve head (ONH) also revealed segmental disc swelling of both eyes.

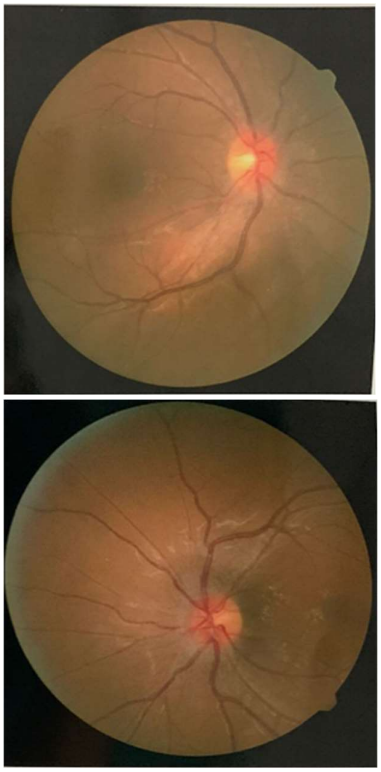

Figure 3. Disc swelling noted on the left eye

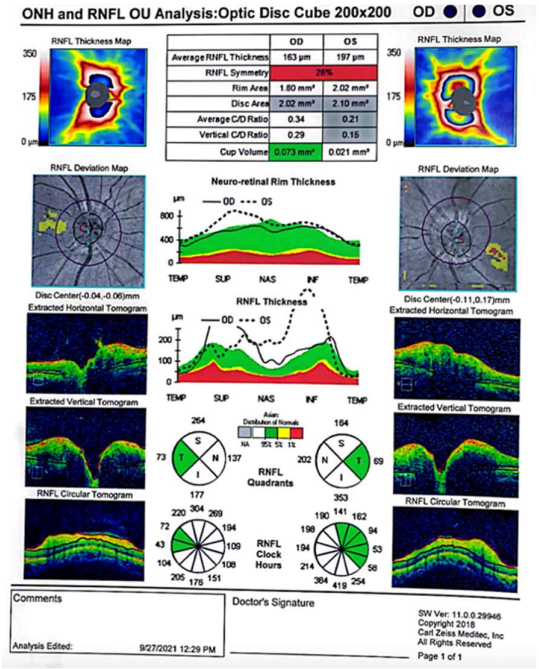

Figure 4. OCT of the Optic Nerve Head showed bilateral segmental disc swelling.

Head and orbit's MRI showed, a solid sinonasal mass was obtained that was characteristic of a malignancy, extending to the optic chiasma and bilateral temporal lobes involving the surrounding muscles destroying the Os. Clivus and Os. Bilateral sphenoid, encompassing the common carotid artery bilaterally and the optic nerve bilaterally. Bilateral maxillary sinusitis was also found. The patient was then consulted to the Otolaryngology Department.

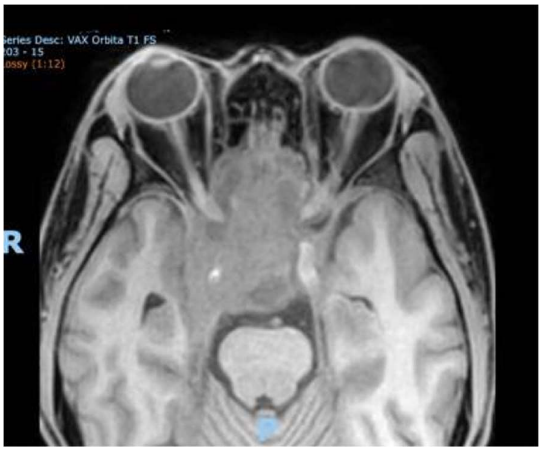

Figure 5. Brain and orbits MRI revealed a malignant solid sinonasal mass, extending to the optic chiasma and bilateral temporal lobe, destroying Os. Clivus and Os. Bilateral sphenoid, encompassing the common carotid artery and optic nerve bilaterally.

From the examination carried out by the Otolaryngology Department, a physical examination was carried out and a mass as high as the concha media was found in the left nasal cavity, so the patient was then diagnosed with a suspected left sinonasal tumor. A biopsy was carried out chronic granulomatic inflammation accompanied by necrosis which suggested a tuberculous process. Then patient was consulted to the pulmonology department for further examination.

From the results of the patient's chest X-ray, there were no abnormalities, and the results of the microbial rapid test performed by the pulmonologist did not detect Mycobacterium tuberculosis.

A month later, the patient was scheduled for a repeat biopsy of the left medial turbinate and right stenoidal recess and also a CT scan with contrast by the Otolaryngology Department. From the results of the examination found granulomatic chronic inflammation in the area of necrosis.

Head and orbit's CT-Scan results found a mass including bilateral sphenoidal sinuses, bilateral ethmoidalis, and posterior nasal cavities, which destroyed the walls of the paranasal sinuses, the sella turcica, and extended to the intracranial region sella (attached to the pituitary gland) and the cavernous sinuses covering the bilateral internal carotid arteries. cavernosa with a mass size that tends to be reduced compared to previous MRI examinations.

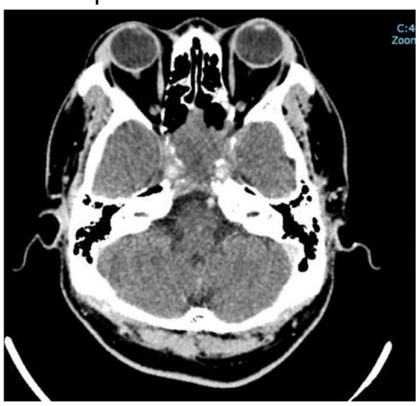

Figure 6. CT Scan results 4 months after first visit, the size of the mass tended to be reduced compared to the initial MRI.

Visual acuity and ophthalmoplegia remained the same since the last visit, but the optic nerve head progressed to atrophic on both eye.

DISCUSSION

Proptosis and ophthalmoplegia may be caused by many conditions such as infectious disease, vascular conditions, inflammatory conditions or certain types of neoplasm that invade the cavernous sinuses, orbital tissue, or the lacrimal gland. [3,4]

Sinonasal tumors are tumors that occur around the nasal cavity or paranasal sinuses. The paranasal sinuses themselves consist of four pairs of sinuses lined with pseudostratified columnar epithelium. The first is the maxillary sinus, which is located under the eye in the maxillary bone, the frontal sinus, above the eye in the frontal bone, the ethmoid sinus, formed of several air cells within the ethmoid bone between the nose and eye and the sphenoid sinus, located in the sphenoid bone body. [5,6]

There are many causes of masses in the sinuses and paranasal sinuses, both neoplastic and non-neoplastic lesions. Polyps are the most common cause of nasal obstruction in adults with a prevalence of approximately 4% in the general population. Sinonasal masses are often found in the ENT field, usually the patient will experience olfactory disturbances, nasal discharge, and epistaxis. If the disorder worsens, eye symptoms such as decreased visual acuity, epiphora, diplopia or exophthalmos can occur. As experienced by this patient, initially the patient complained that there was a stuffy nose and yellowish discharge, then not long after that the patient complained of visual acuity which was getting worse. The patient also had bilateral proptosis. [7-9]

Decreasing visual acuity in this patient might be caused by optic nerve compression in the sinonasal tumor. Optic nerve compression in sinonasal tumours usually occurs at the intra-orbital portion of the optic nerve, but it may also involve the intracranial portion and optic chiasm in cases of aggressive tumours. Compressive optic neuropathy can occur by compressing the vascular supply and causing ischemia to the nerve or directly causing mass effect upon the axons, thereby impairing axonal transport and signal transmission. [10,11]

Ophthalmoplegia in this patient may be caused by tumor invasion into the cavernous sinus area as shown in the patient's last MRI results. The cavernous sinuses are part of the dural venous sinuses of the brain which contain many nerve vessels. The cavernous sinus lies in the sella turcica and extends from the anterior superior orbital fissure to the petrous portion of the posterior temporal bone and is about 1 cm wide and 2 cm long. There are cranial nerves located in the cavernous sinus, namely the oculomotor nerve (CN III), trochlear nerve (CN IV), ophthalmic nerve (V1), maxillary nerve (V2), abducens nerve (CN VI), and the sympathetic plexus around the internal carotid artery. According to Ebert et al, there

are about 5% of ophthalmoplegia cases due to cavernous sinus syndrome that affects CN III, CN IV, and CN VI, for example, caused by a tumor or inflammatory process. Tumors in the cavernous sinus may be pituitary adenomas, meningioma, schwannomas, lymphomas, perineural tumor metastases, or direct invasion of the tumor (as in naso-pharyngeal carcinoma). Pituitary adenoma is the most common tumor-causing lesion involving the cavernous sinus. Lesions to the cavernous sinus or orbit can cause simultaneous lesions of cranial nerves III, IV, and VI as well as sympathetic nerves involving the iris, causing total ophthalmoplegia. Involvement of cranial nerves V1 and V2 can cause facial sensory loss in the upper and middle parts, but in patients there is no sensory loss on the face so it can be ascertained that there is no involvement of cranial nerves V1 and V28. [12,13]

Most sinonasal neoplasms have non-specific imaging features. The effect on adjacent bony structures may lead to a benign lesion in the presence of pressure erosion changes, to a malignant lesion if there is lytic damage or to sarcoma if there is a periosteal reaction. [14]

In a study conducted by Chu et al in 2012, most of the causes of sinonasal carcinoma are squamous cell carcinoma, which is associated with risk factors such as alcohol and smoking. In this patient smoking risk factors were found but the results of the biopsy showed chronic granulomatic inflammation accompanied by necrosis which suggested a tuberculous process. Sinonasal tuberculosis is a very rare condition. The diagnosis of sinonasal tuberculosis can only be established based on the following criteria: (a) no clinical response to antibiotics (b) presence of caseous granulomatous inflammation on histopathological analysis and (c) identification of Mycobacterium tuberculosis in the specimen.

Tuberculosis of the eye may produce symptoms of proptosis because of an existing mass or diplopia resulting from extraocular muscle involvement. Also reported in cases of orbital tuberculosis due to spread from paranasal sinuses, globe dystopia, ophthalmoplegia, loss of visual acuity, epistaxis, and contralateral cutaneous-maxillary sinus fistula. Involvement of the frontal sinus, sphenoid, and zygoma has also been reported in cases of extrapulmonary tuberculosis. On histological examination, epithelioid granulomas with Langharns' giant cells and caseation necrosis were found. The diagnosis is then established by the presence of Mycobacterium tuberculosis DNA through polymerase chain reaction (PCR) examination. While the patient did not find Langharns' giant cells and the polymerase chain reaction (PCR) test had not been carried out, so the diagnosis of sinonasal tuberculosis with orbital involvement could not be established. [15-18]

CONCLUSION

Cases of bilateral ophthalmoplegia and visual loss caused by sinonasal tumours are due to their extension to the cavernous sinus resulting in multiple cranial nerve paresis and optic nerve compression. Holistic approaches with neuroimaging such as MRI and CT scans are needed to determine the location of the lesion and the possible etiology, so that further treatment can be decided.

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