An Optic Nerve Sheath Meningioma Presenting with Choroidal Folds and Vision Loss

Koroid Katlantıları ve Görme Kaybı ile Seyreden Optik Sinir Kılıfı Menenjiomu

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ABSTRACT

We report an optic nerve sheath meningioma case in a 57-year-old woman admitted to our clinic with unilateral vision loss. Best corrected visual acuity (VA) was 1/10 on the Snellen chart at admittance. The only fundoscopic finding was macular choroidal folds. The orbital MRI scan revealed optic nerve sheath meningioma. The patient was referred to the neurosurgery department and received radiotherapy. Six months after treatment VA was 2/10 in the right eye and the choroidal folds persisted on fundoscopy. Moreover, based on this case we report the clinical course and treatment options for optic nerve sheath meningioma.

Key Words: Optic nerve sheath meningioma, choroidal folds.

ÖZ

Kliniğimize tek taraflı görme kaybı şikayetiyle başvuran 57 yaşında bayan hastada saptanan optik sinir kılıfı menenjiomu olgusunu sunduk. Başlangıçta hastanın en iyi düzeltilmiş görme keskinliği Snellen eşeli ile 1/10 idi. Fundus muayenesindeki tek bulgu koroidal katlantılardı. Orbital MR sonucunda optik sınır kılıfı menenjiomu saptandı. Hasta beyin cerrahisi kliniği ile konsülte edildi ve radyoterapi uygulandı. Tedaviden 6 ay sonra sağ gözde görme keskinliği 2/10 idi ve fundus muayenesinde koroidal katlantılar devam etmekteydi. Bu olgunun ışığında optik sinir kılıfı menenjiomunun klinik seyri ve tedavi seçeneklerini de değerlendirdik.

Anahtar Kelimeler: Optik sinir kılıfı menenjiomu, koroidal katlantılar.

INTRODUCTION

Optic nerve sheath meningioma (ONSM) is a benign neoplasm that arises from the meningothelial cap cells of arachnoid villi. It may develop anywhere along the course of the optic nerve, from globe to prechiasmal intracisternal optic nerve. Slowly progressive unilateral visual loss is the most common complaint encountered and proptosis, relative afferent pupil defect, dyschromatopsia, and eye lid edema may also be present. At presentation, the optic nerve head may appear normal, atrophic, or swollen and optociliary shunt vessels may be visible. Imaging characteristics are usually sufficient to allow diagnosis of ONSMs. Both CT and MRI show diffuse tubular enlargement of the optic nerve. 1-3

We report an ONSM patient that presented with choroidal folds and give the results of some available treatment options for ONSM.

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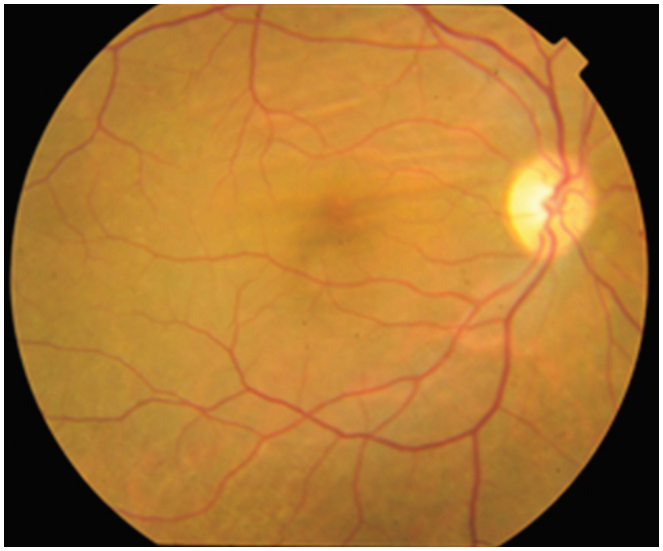

Figure 1: *Right eye fundus photography shows macular choroidal folds.*

CASE REPORT

A 57-year-old woman was admitted to our clinic with unilateral vision loss in the right eye. Best corrected visual acuity (VA) was 1/10 in the right eye and 10/10 in the left on the Snellen chart. Intraocular pressures were within normal limits and there was no pathologic sign upon anterior segment examination.

Pupillary reflex was bilaterally normal. At fundoscopic examination, choroidal folds were observed in the macula and the optic nerve seemed to be normal (Figure 1) in the right eye. Moreover, choroidal folds were seen on the OCT scan (Figure 2). The orbital MRI scan revealed tubular enlargement of the right optic nerve (Figure 3), and so the patient was referred

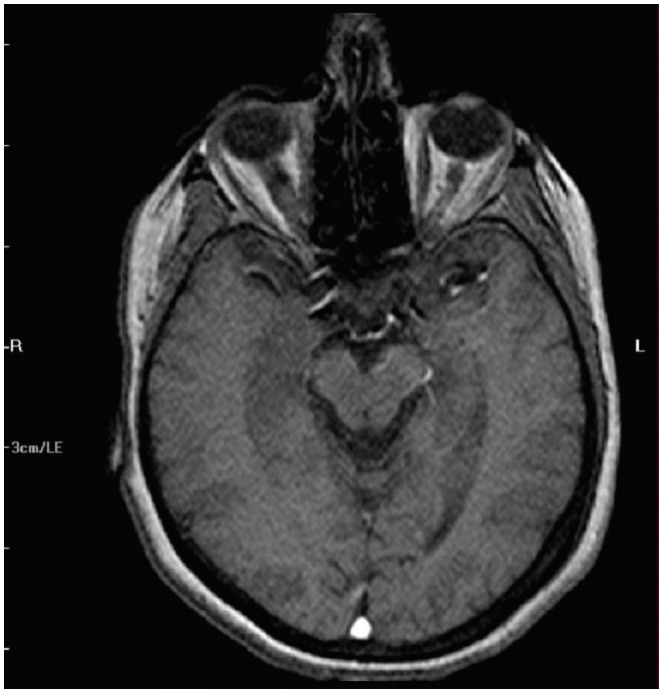

Figure 3: *The axial T1-weighted MRI scan image of the patient shows a right hypointense enlargement of the optic nerve.*

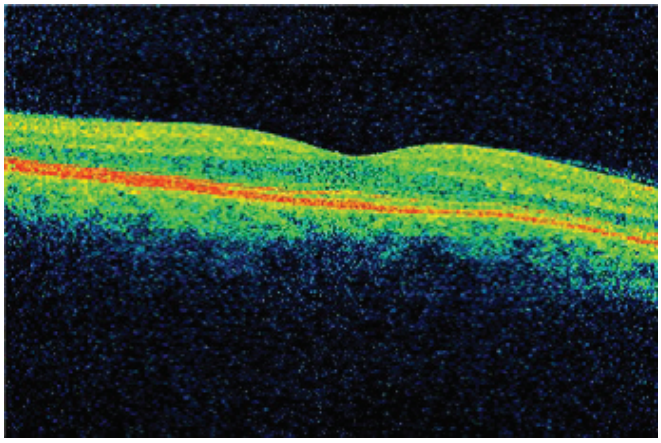

Figure 2: *OCT scan of right eye demonstrates choroidal folds.*

to the neurosurgery department with a prediagnosis of right ONSM. After confirmation of the diagnosis, the patient received a total dose of 50 Gy radiotherapy fractioned over the course of 6 weeks. Six months after treatment VA was 2/10 in the right eye. The choroidal folds persisted on fundoscopy.

DISCUSSION

Optic nerve sheath meningiomas are the second most common type of optic nerve tumors. They usually occur in middle-aged women. Mean age at presentation is 40.8 years overall, varying from 42.6 years for women and 36.1 years for men.[1-5] To our knowledge, the youngest ONSM patient was a 2.5-year-old child.[6] As a rare form, ONSM may represent bilaterally and usually is related to Neurofibromatosis type 2. The most common symptom is a gradual, painless, unilateral vision loss, while optic nerve head changes generally accompany the symptoms.

Choroidal folds have been associated with a myriad of conditions including axial hyperopia, hypotony, uveal effusion, idiopathic intracranial hypertension, orbital tumors, central serous chorioretinopathy, and posterior scleritis.[7,8]

In this patient, choroidal folds were the only fundoscopic finding and these may contribute to vision loss. The choroidal fold formation was probably due to stretching of the optic nerve.

Radiologic scans are the keystone of the diagnosis and MRI fat suppression and CT scans should be performed on the way through. ONSMs are characterized by classification on a histologic basis. Even though CT scan is a better technique to demonstrate this pattern, the MRI scan is still the first choice modality to diagnose ONSM. ONSMs are typically isointense or slightly hypointense to brain and optic nerve tissue on T1 weighted images and hyperintense (may also be hypointense) on T2 weighted images.

After administration of intravenous contrast, a central linear lucency is left inside the enlarged optic nerve; this is called the "tram track" sign. The traditional treatment approach to ONSMs was conservative, including regular automated perimetries and MRI scans. Despite being a slowly progressive tumor, surgery is still an option in patients who have intracanalicular or intracranial extension of the tumor. On the other hand, these tumors are not associated with any mortality or significant neurological morbidity and rarely spread. Lindblom et al.,[9] reviewed a large series of patients who underwent radical tumor resection of cranio-orbital meningiomas and reported improved visual function in 27%, stable visual function in 62%, and worsening vision in 11%.

Turbin et al.,[10] studied 59 patients with better than no light perception at presentation. The patients were divided into 4 groups: 13 patients (Group 1) were observed only, 12 patients (Group 2) had surgery only (4 biopsy or partial resection, 8 total resection), 18 patients (Group 3) received radiation alone, and 16 (Group 4) had surgery and radiation (14 patients in this group had biopsy or partial resection and radiation, and 2 had total resection and radiation). All of the treatment groups showed statistically significant visual loss except the radiation-only group, which showed a trend for loss that was not statistically significant. Pitz et al.,[11] reported the results of treatment of 15 patients with ONSMs using a more precise form of fractionated radiation therapy-stereotactic fractionated conformal irradiation. Pitz et al.,[11] found that over a mean follow-up period of 37 months after completion of treatment, there was no evidence of tumor growth or extension, none of the treated seeing eyes experienced deterioration of vision, and several eyes improved with respect to visual acuity, visual field, or both. In addition, none of the radiated eyes developed radiation retinopathy or optic neuropathy, and none of the contralateral eyes showed evidence of radiation injury. Richards et al.,[12] reported similar findings, with a definite visual improvement and no severe complication. The standard radiation dose for ONSM is 50 to 54 Gy, administered in fractions of 1.7 to 1.8 Gy over 6 weeks.[13] This protocol was derived from retrospective studies of progression-free survival rates in patients with benign intracranial meningioma in all locations.[14]

Although meningiomas are known to be hormone therapy sensitive tumors, clinical studies using tamoxifen and mifepristone have been disappointing.[15] In a case report, Paus et al. administered 20 mg/kg hydroxyurea treatment to a 46-year-old woman with ONSM. Visual acuity was 0.05 at onset and improved to 0.8 after 10 months' follow up. However, no change in tumor size was observed by MRI.[16]

In conclusion, optic nerve sheath menengioma should be kept in mind in patients with vision loss. Despite the slow progression pattern, rare conditions, such as intracranial spreading, prompt physicians to search for new treatment modalities. Developing radiologic techniques show promise for the future of ONSM treatment and hydroxyurea may be a reasonable therapeutic alternative to radiotherapy; however, large series are necessary to evaluate the safety and effectiveness of this modality.

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