Primary Central Nervous System Lymphoma of Optic Chiasma: Endoscopic Endonasal Treatment

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Background: Isolated primary central nervous system lymphoma arising from anterior visual pathway is very rare.

Patient Presentation: A 76-year-old immunocompetent previously healthy man presented bilateral decreased visual acuity in 1 month. Pituitary magnetic resonans imaging (MRI) showed a lobulated mass with homogeneous enhancement after gadolinium administration that arising from optic chiasm suggested that inflammatory disease or an optic glioma. The patient underwent an extended endoscopic endonasal transsphenoidal surgery. Postoperative course and outcomes were wonderful. Histopathological diagnosis was diffuse large B-cell lymphoma. The patient underwent investigations for systemic lymphomatous involvement, did not detect any evidence of systemic disease.

Conclusion: In this case, we claimed that differential diagnoses of anterior visual pathway lesions are difficult because of similarity of lesions on clinical and radiological examinations. Biopsy is essential for these lesions. As a biopsy technique, endoscopic endonasal transsphenoidal approach is safer and more effective than open procedures.

Key Words: endonasal, endoscopic, lymphoma, optic chiasm, visual acuity

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Primary central nervous system lymphoma (PCNSL) has seen progressively increased number in last 30 years.1 However, isolated PCNSL of the optic tract, chiasm, and optic nerve is extremely rare1,2 and in the absence of coinciding systemic lymphomatous involvement, diagnosis is difficult because of similarity with neoplastic, inflammatory, or autoimmune infiltrations on clinical and radiologic manifestations. In this article, we present an immunocompetent patient with histologically proven PCNSL arising from optic chiasma that operated with endoscopic endonasal transsphenoidal approach.
(1.06 mIU/mL), luteinizing hormone (<0.2 mIU/mL), total testosterone (<0.1 ng/mL), and morning cortisol level (0.96 μg/dL) were low. Prolactin, growth hormone, and insulin-like growth factor-1 levels were normal. Complete blood count and erythrocyte sedimentation rate were normal and enzyme-linked immunosorbent assay for human immunodeficiency virus was negative.

Because of repetitive syncope attacks and progressive visual deterioration, the surgery planned for the patient as pure endoscopic endonasal transtuberculum approach. The patient underwent surgery via binostril approach, using 0- and 30-degree rigid endoscopes (Karl Storz, Tutlingen, Germany). The extended transtuberculum approach was used. Under the intact dura matter, yellow-white, solid tumor was seen and frozen section biopsy was done. The result of frozen section came as malignant tumor after that the tumor was resected subtotally included suprasellar part of tumor. Optic chiasma and tractus superior and anteriorly, bilateral carotid arteries laterally, normal hypophysis inferiorly, and third ventricule posteriorly were used as margins of resection (Fig. 3).

After resection, the dura defect was reconstructed with free fat, tensor fascia lata autografts, and fibrin tissue adhesives, using multilayer skull base reconstruction technique. At the end, nasal Foley catheter was placed for immobilization of reconstruction materials. External lumber drainage catheter was replaced. There is no perioperative or postoperative complication.

Early postoperative pituitary MRI showed that the tumor was excised subtotally (Fig. 4). After surgery, the patient did not suffer additional visual acuity or visual deterioration. The patient remained in a state of panhypopituitarism after the operation and was given hormone replacement therapy. Histopathology of tumor showed aggressive pleomorphic lymphoid atypical cellular infiltration with high proliferation rate and increased apoptosis. Immunohistochemical staining atypical cells were positive with CD20, Bcl-6, and MUM-1. Ki-67 proliferation index was 60% (Fig. 5). These findings with the radiology and other data addressed a PCNSL.

The patient was referred to the department of Hematology for further investigations of systemic lymphomatous involvement, but there was not detected any systemic involvement. The patient underwent chemotherapeutic treatment for PCNSL.

**DISCUSSION**

In this study, we presented an immunocompetent, visually deteriorated patient who had PCNSL arising from optic chiasm that was proved with preoperative MRI scans, postoperative pathological examination, and further examinations for systemic lymphomatous disease. Although the incidence of PCNSL has been progressively increased in the last 3 decades, isolated PCNSL of the anterior visual pathway is still rare. Lymphomatous involvement of anterior visual pathway usually occurs as a result of metastatic spread of systemic non-Hodgkin lymphoma. There was an isolated PCNSL of the optic nerve reported by Kansu et al that resected optic nerve for diagnosis by craniotomy.

Differential diagnosis of anterior visual pathway lesions is difficult, and range is very wide that includes inflammatory, autoimmune, or neoplastic infiltrations, for example, optic glioma. Because early diagnosis and treatment are important for PCNSL, the patients who have isolated anterior visual pathway lesions
without a specific diagnosis should be considered for biopsy. For the review of the literature, there have been 8 patients including our patient, that localized biopsy was performed on the anterior visual pathway.\(^4\)\(^5\)\(^6\) (Table 1). The mean age of patients was 65 years (range; 49–76) and 5 of 8 patients were men.\(^4\)\(^5\)\(^6\)\(^7\)\(^8\)\(^9\)\(^10\) All patients had visual deterioration as an initial symptom. Homogeneous enhancement of the lesions after gadolinium administration was seen in all except 1 patient in preoperative MRI scans. Seven of 8 patients, except our patient, underwent open biopsy for diagnosis. We performed biopsy with endoscopic endonasal transtubercular approach. In this aspect, our case is unique. In isolated anterior visual pathway lesions, maintaining visual function is very important. Providing direct access to the lesion and not requiring optic nerve excetration, endonasal endoscopic technique is superior for the preservation of visual function compared to open surgery. Because endonasal endoscopic technique provides better visualization, illumination and lesser morbidity as compared to open procedures, endonasal endoscopic procedure should be considered instead of open surgery.

### REFERENCES


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