Suprasellar Meningioma Simulating Atypical Retrobulbar Optic Neuritis: A Diagnostic Dilemma For Neuro-Ophthalmologists

M Kamble, P Subramanian, R Juneja, P Sune, S Dulani, P Tidake, S Acharya, P Kalakoti, Z Quazi

Abstract

Atypical optic neuritis (ON) represents a frequent clinical situation in neurology and ophthalmology. Optic neuritis typically affects young adults ranging from 18–45 years of age, with a mean age of 30–35 years. There is a strong female predominance. The annual incidence is approximately 5/100,000, with a prevalence estimated to be 115/100,000 but very rarely can also occur in males and in any age group. [1]

Suprasellar meningioma as a cause of atypical retrobulbar optic neuritis leading to sudden vision loss is rare. It is usually asymptomatic and often remains undiagnosed. We report a very unusual case of Suprasellar Meningioma in a 55 years old male, presenting as atypical retrobulbar optic neuritis.

INTRODUCTION

Optic neuritis is an acute inflammatory disease. The disease is characterized by unilateral sudden loss of vision in the affected eye, often accompanied by periorcular pain. Majority of cases are idiopathic in nature.

Retrobulbar Optic Neuritis which presents as normal appearance of Optic disc is found in 65% of cases. The visual defects seen are usually general depression, altitudinal field loss or central scotoma. Visual acuity is regained to near normal in most of the typical cases. [2] In Asian people, the incidence of multiple sclerosis as a cause of Optic neuritis is very low. [3]

Suprasellar Meningiomas are generally benign, well circumscribed, slow growing, asymptomatic and often remain undiagnosed. [4] Due to their close proximity with vital neurological structures, the treating physicians have a considerable difficulty.

CASE REPORT

A 55-year-old male came to our outpatient department with the chief complaint of sudden onset diminution of vision of his right eye since 2 months and gradual diminution of vision in left eye since few months. He was treated elsewhere with oral prednisolone 2 months before this visit, and the vision of the right eye improved then. At visit, the visual acuity of the right eye was 6/24, and the left 6/12. The anterior segment was quiet bilaterally except for few early cataractous changes in lens. The fundus examination revealed bitemporal optic disc pallor with few soft small drusens around the macula, OU (Fig1a,b). The disc showed clear margin, pinkish color and no elevation, OU. The first impression was of dry AMD OU with sequel of central serous chorioretinopathy OD.
Two weeks later, the patient came back with further decrease in the vision of his right eye. The visual acuity was 6/24 in his left eye still, but the vision of the right eye went down to "counting fingers" only. Painful sensation on eye–ball movement was also noted around the right eye. There was no proptosis bilaterally. The ocular examination remained nothing particular. There was a marked RAPD sign in his right eye. The color vision of the left eye was normal, and totally absent in the right eye. The visual field revealed bitemporal hemianopia (Fig 2a,b).
Under the impression of retrobulbar optic neuritis, OU, the patient was treated with methylprednisolone 1g/day intravenously. The vision recovered well and rapidly. Three days later, the visual acuity improved to 6/12 OU, and the color vision improved to 12/21 on Ishihara color testing plate. So he was discharged with oral prednisolone 60 mg/day.

One week later, he noted exacerbation of the right eye vision again. The visual acuity was again 6/60 OD and 6/18 OS. We increased the dosage of prednisolone to 50 mg/day, but even after 2 weeks, the visual acuity did not show any improvement. During that time, the ocular examination remained unremarkable and he was admitted again for further survey.

Brain MRI was done, and a 3 x 2.5 x 2.5 cm tumor mass was found in the planum sphenoida and tuberculum sella region, with compression of bilateral optic nerves (Fig. 3). So, he was sent to the neurosurgical department for management. A left craniotomy with removal of the tumor mass with duraplasty and reconstruction of the skull base was done smoothly. The pathological report was meningioma, meningotheliomatous type.

After removal of the tumor, the visual acuity improved to 6/12 OD and 6/9 OS with spectacles.

**DISCUSSION**

Optic neuritis is diagnosed based on clinical symptoms and signs, so it is not rare to misdiagnose a disease as an optic neuritis, especially the retrobulbar type. In this case, there were several clues that were misleading. First of all, there was no proptosis in appearance, no headache from history, and the visual loss was relatively quick compared with the "traditional image" of a brain tumor. These did not lead us to an impression of a brain tumor.

Compressive optic neuropathy caused by a suprasellar tumor is not rare at all. Generally speaking, there are signs such as proptosis, disc swelling, headache, progressively deterioration of visual symptoms, or other neurological signs. These were not obviously found in this particular patient, who finally was proved to be a case of compressive optic neuropathy caused by a suprasellar meningioma.

Another interesting point in this case was the rapid and excellent response to intravenous methylprednisolone, which
is also seen in optic neuritis cases. However, this condition can happen in some infiltrative disorders like leukemia, but not so often in a solid-mass lesion like meningioma. Prednisolone might reduce the edema surrounding a tumor, and alleviate the compressive effect. Usually, it takes a long time for a brain tumor to cause visual disturbance, and the victim usually perceives persistent and progressive visual dysfunction instead of sudden visual loss or fluctuation of visual functions, but these were what this patient showed. In this case, it seemed that the tumor as well as the perifocal edema extending from the meningioma produced the compressive effect on the optic nerve, the latter can be alleviated by steroid effect, however, the former only by surgical removal.

It seems that a neuroimaging study in every case is the best tool to avoid misdiagnosis. However, the ONTT concluded that neuroimaging is of limited practicality and is not cost-effective in the diagnosis of optic neuritis. In the rare cases of a compressive lesion masquerading as optic neuritis, the patients’ atypical courses will alert the clinician to the need for neuroimaging studies. That was exactly what happened in this case, and a delay in diagnosis and treatment seemed unavoidable. To our regret, the right eye vision once returned to 6/6 but ended with “hand motion” only. It might have been better if the diagnosis and management had not been delayed.

In an analysis of blindness from intracranial tumors in 60 cases, Zhou et al. [5] found 61.7% of blinding tumors located in the sellar region in 60 cases. Most of them were pituitary adenoma (22 out of the 60), and only 4 were meningioma. The ocular findings showed papilledema in 17 cases (28.3%), and optic atrophy in 40 cases (66.7%).

Monocular blindness was noted in 37 patients; of them 10 showed decreased vision in the other eye also. Bilateral blindness was noted in 23 patients. It may be due to the conservative personality and tradition in Chinese people that a delay of seeking medical help usually presents. So, there is such a high ratio of blindness as well as high percentage of optic atrophy. Additionally, of the 60 patients, 5 had been diagnosed previou-sly as optic neuritis, retrobulbar optic neuritis, or optic nerve ischemic lesions.

Symon and Rosenstein reported that in 101 patients with suprasellar meningiomas, loss of vision was noted as the first symptom in 74.2%. [6] The Ophthalmologists may be the first physicians they consulted. If we could find the tumor earlier, the patients might not go completely blind. Rosenstein and Symon reported that 41.8% of patients who complained of monocular visual loss also showed some vision reduction of the other eye. [7] This may be an important clue to the ophthalmologists for the need of a neuroimaging study.

The experience of Jallu et al [8] in Saudi Arabia is more fascinating. They reported 70 patients with suprasellar meningioma, and in 80% of them, the tumors were larger than 5 cm in diameter. Sixty patients showed deterioration of vision as the primary symptom. Thirty-four of them also showed headache before visit. The correlation of vision loss and intracranial tumor is close, especially in suprasellar tumors. In this report, 31 patients showed no light perception in one or both eyes, and bilateral optic atrophy was noted in 45 patients. This also might be due to a delay in seeking medical help. It’s a tragedy that so many people went blind because of a disease that could have been treated earlier.

CONCLUSION

In an Asian Country like India, the cases of Multiple sclerosis are scanty unlike the Western world. So, usually MRI is not performed in each and every case of Optic Neuritis. Its diagnosis is made on the basis of its clinical features. The visual prognosis in patients with suprasellar meningioma who undergo excision of tumor is excellent. But recurrences tend to occur even after a period of 10 years following surgery, so long term follow-up is essential. However, in all cases of Optic Neuritis, neuro-imaging must be carried out as a mandatory investigation to rule any brain tumor or other conditions causing compressive optic neuropathy must be ruled out.

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STUDY APPROVAL

The study was conducted at Acharya Vinoba Bhave Rural Hospital during January 2013 to March 2013. It was submitted and approved by Institutional Ethical Committee, Acharya Vinoba Bhave Rural Hospital, Datta Meghe Institute of Medical Sciences.

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Author Information

Mala Kamble
Department of Ophthalmology, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India
rakeshjuneja86@gmail.com

Prem Subramanian
Neuro-ophthalmology Division, The Wilmer Eye Institute, Johns Hopkins University School of Medicine
Baltimore, Maryland, USA

Rakesh Juneja
Department of Ophthalmology, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India

Pradeep Sune
Department of Ophthalmology, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India

Somya Dulani
Department of Ophthalmology, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India

Pravin Tidake
Department of Ophthalmology, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India

Sourya Acharya
Department of Ophthalmology, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India

Piyush Kalakoti
Rural Medical College, Pravara Institute of Medical Sciences
India

Zahiruddin Sayeed Quazi
Department of Community Medicine, Acharya Vinoba Bhave Rural Hospital
Sawangi, Wardha, India