Recurrent abducens nerve palsy with optic perineuritis

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Dear Sir,

I am Joo Yeon Kim, from the department of Ophthalmology of Kim's Eye Hospital in Seoul, Korea. I write to present a case report of recurrent abducens nerve palsy with optic perineuritis. Abducens nerve palsy is associated with trauma, viral infection or inoculation, central nervous system tumors, elevated intracranial pressure and idiopathic cause. Optic perineuritis is an uncommon noninfectious inflammation of the optic nerve sheath and perineurial fat. Optic perineuritis presents as an orbital apex syndrome with optic neuropathy, proptosis, and ophthalmoplegia. It occasionally occurs as a manifestation of a specific infectious or inflammatory disorder. We report a case of abducens nerve palsy with visual disturbance, which was associated with optic perineuritis confirmed by magnetic resonance imaging (MRI). A 59-year-old woman developed sudden visual disturbance and limitation of abduction in the left eye simultaneously. She also complained of pain on eyeball movement, especially when trying to abduct. She was otherwise healthy and had no specific ocular history. We full ophthalmologic examination were performed including best corrected visual acuity, color vision, visual field test by Humphrey 24-2 automated perimetry (Zeiss, Germany), fundus examination, relative afferent pupillary reflex and ocular motility. Laboratory tests were performed including complete blood cell count, the erythrocyte sedimentation rate, C-reactive protein, rheumatoid factor, anti-nuclear antibodies, anti-neutrophil cytoplasm antibodies, and serologic tests such as syphilis, Toxoplasma, Cytomegalovirus, Varicella zoster virus, Hepatitis-B virus, Hepatitis-C virus, and Human immunodeficiency virus. We also conducted chest x-ray, and magnetic resonance imaging study. The best corrected visual acuity was 0.2 in the left eye. Total dyschromatopsia was noted, and visual field test disclosed an inferior arcuate scotoma. On fundus examination, there was no disc swelling, nor were there splinter hemorrhages. A marked left relative afferent pupillary reflex was present. The degree of abduction limitation was -4. Laboratory tests were normal, as was the chest x-ray. MRI showed increased fluid collection in the subarachnoid space surrounding the left optic nerve (Figure 1A). The diagnosis of optic perineuritis with abducens nerve palsy was made. Steroid pulse therapy was started: methylprednisolone 1 g/day IV for 3 days, 40 mg/day PO for 7 days, and tapered over 1 month. Three days after the implementation of steroid pulse therapy, the color vision fully recovered, and the inferior arcuate scotoma disappeared. After 11 days, the best-corrected visual acuity was improved to 1.0, and the degree of abduction limitation decreased to -0.5. No relative afferent pupillary reflex was seen. The abducens nerve palsy completely resolved 3 weeks after steroid pulse therapy. At 9 months after the first attack, limitation of abduction in right eye and right optic perineuritis was developed (Figure 1B). Repeat steroid pulse therapy was administered and limitation of abduction was resolved at 10 days after the pulse therapy. At her last visit, 3 months after the second attack, the patient's best-corrected visual acuity was 1.0, the relative afferent pupillary reflex was normal, and there was no limitation of abduction.

Berlit et al[5] evaluated 165 patients with abducens nerve palsy and noted the following distribution of etiologies: vascular origin (29.7%), inflammatory disease (19.4%), tumor (10.9%) and trauma (3.1%). The condition can also be idiopathic. In our case, the etiology thought to be idiopathic or viral infection which is undetected. Abducens nerve palsy is rarely associated with optic perineuritis. Purvin et al[6] reported that 2 of 14 examined patients had slight abduction deficits with optic perineuritis. However, abducens nerve palsy with optic perineuritis has never been reported before.

Optic perineuritis is a term used to describe an orbital inflammatory disease in which the main focus is the optic
Based on clinical features, optic perineuritis is likely to be misdiagnosed as optic neuritis. Purvin et al. suggested several features to distinguish optic neuritis from optic perineuritis based on the data from 14 patients with visual loss, eyeball pain, or both. Compared with optic neuritis, optic perineuritis develops in a broader age group, progresses over weeks, and responds more promptly and dramatically to corticosteroid treatment. MRI is now the preferred diagnostic modality, since the current MRI protocols of fat suppression in conjunction with Gd-DTPA contrast enhancement provide the best images of the optic nerve sheath, as well as the optic nerve. Imaging of optic perineuritis shows a typical pattern of enhancement around the optic nerve. In the present patient, we noted increased fluid collection in the subarachnoid space surrounding the left optic nerve; the so-called "tramtrack" appearance on axial view and "doughnut" appearance on coronal view.

The effect of steroid therapy was reported in optic perineuritis and 14 patients with optic perineuritis demonstrated dramatic relief of pain and visual loss within hours, and the entire study population demonstrated symptom resolution within a day. In our patient, the dramatic effect of steroid therapy was seen not just on optic perineuritis, but also on abducens nerve palsy.

REFERENCES


