Idiopathic inflammatory perioptic neuritis is a rare inflammatory condition of the optic nerve sheaths. The differential diagnosis is important because the disease has radiologic findings resembling those of optic nerve sheath meningioma and management is quite different in each condition. A case of bilateral idiopathic inflammatory perioptic neuritis is reported in which the computed tomographic (CT) appearance simulated optic nerve sheath meningioma and there was progressive visual loss.

CASE REPORT

Bilateral Idiopathic Inflammatory Perioptic Neuritis Simulating Optic Nerve Sheath Meningioma on Computed Tomography

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Idiopathic inflammatory perioptic neuritis is a rare inflammatory condition that typically presents with recurrent episodes of visual loss associated with pain, and its neuroradiologic features may resemble those of an optic nerve sheath meningioma. The term idiopathic inflammatory perioptic neuritis has been used to describe a group of inflammatory conditions of unknown etiology affecting the optic nerve sheaths (pia, arachnoid, and dura). We present a case of bilateral idiopathic inflammatory perioptic neuritis whose computed tomographic (CT) appearance mimicked optic nerve sheath meningioma.

Case Report

A 49-year-old woman was referred to the Ankara Numune Hospital Eye Clinic in Ankara, Turkey, for evaluation of progressive visual loss and periorbital pain beginning 2 months earlier. Her visual acuity was 20/70 in the right eye and 20/400 in the left eye. Color vision testing with Ishihara plates revealed bilateral marked dyschromatopsia. A left afferent pupillary defect was present. Eye movements were free but painful for each direction bilaterally; no proptosis was measured. Results of slit-lamp biomicroscopy and oph...

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Presented as a scientific poster at the XIXth Congress of the European Society of Ophthalmology, Hungary, Budapest, June 1-5, 1997.
thalamoscopy were normal. Intraocular pressures were within normal ranges in both eyes. Visual field examination revealed a dense central scotoma bilaterally.

Results of the complete blood cell count and routine biochemical laboratory tests were normal. Erythrocyte sedimentation rate was 40 mm/h, and test results for VDRL, rheumatoid factor, and antinuclear antibody were negative. Serum protein electrophoresis and routine thyroid function tests were within normal ranges, as were the angiotensin-converting enzyme levels. Chest x-rays were reported as normal.

The patient’s CT scan with contrast media revealed thickening in both optic nerves in a tubular and tortuous fashion in the axial plane with a 2-mm section thickness (Fig 1). Visual acuity was hand motions in the right eye and light perception in the left eye. Despite the patient’s age, sex, progressive visual loss, and CT features resembling optic nerve sheath meningioma, her rapidly progressing, painful visual loss and the thickened appearance of the optic nerve could also be interpreted as bilateral inflammatory periorbital neuritis. Magnetic resonance imaging (MRI) was not available, and biopsy could not be performed. Systemic corticosteroid treatment (methylprednisolone, 80 mg/day) was initiated.

One week later her visual acuity was recorded as 20/25 in the right eye and 20/20 in the left eye. Color vision in the right and left eyes were 6/12 and 2/12, respectively as measured by Ishihara plates. A repeated CT scan with the same parameters revealed regression in the thickening and tortuosity of both optic nerves (Fig 2). The clinical and neuroradiologic response to corticosteroid treatment supported the diagnosis of idiopathic inflammatory periorbital neuritis. The corticosteroid dosage was slowly tapered before discontinuation.

One month later, visual acuity was 20/20 in the right eye and 20/30 in the left eye, while color vision was 10/12 and 2/12 in the left and right eye, respectively. The right optic disc was normal, and the left optic disc had temporal pallor. The patient has been followed-up in 6-month intervals for 3 years, and her condition remains unchanged.

Discussion

Optic nerve and sheath enlargement may be the result of various neoplastic and nonneoplastic diseases. Among the neoplastic conditions, optic nerve gliomas, optic nerve sheath meningiomas, neurofibromas, hemangioblastomas, metastases, and leukemias are mentioned. Some nonneoplastic causes leading to the clinical pattern are increased intracranial pressure, optic neuritis, dysthyroid orbitopathy, granulomatous optic neuropathies (especially sarcoidosis), and traumatic hematomas.

The term idiopathic inflammatory periorbital neuritis was first used in 1985 by Dutton and Anderson although Rush et al previously described a similar case. Dutton and Anderson performed biopsies in 13 cases with visual loss and evidence of optic nerve or sheath enlargement on CT scans. They described 4 patients in whom adequate histologic samplings failed to demonstrate meningioma and there was presence only of inflammatory infiltration of dural sheaths and edematous or dense fibrous tissue. We found a total of 9 cases of idiopathic inflammatory periorbital neuritis reported in the literature: 2 of them bilateral and 7 unilateral.

Idiopathic inflammation of the optic nerve sheaths presents as an enlargement of the optic nerve and sheaths on CT scans. The enlargement may be fusiform, but the radiographic pattern is usually a uniform, tubular, and diffuse thickening. Although MRI using gadolinium and fat-suppression techniques has been reported to be effective in differentiating the optic nerve sheath meningiomas from optic nerve gliomas, differentiating idiopathic inflammatory periorbital neuritis from optic nerve sheath meningiomas is not always possible.

The diagnosis of idiopathic inflammatory periorbital neuritis is usually established at biopsy. Pathological changes were described in detail in the electron

![Fig 1.—Orbital computed tomographic scan reveals tubular thickening and tortuosity of the optic nerves.](image1)

![Fig 2.—Computed tomographic scan shows regression in thickening and tortuosity of the optic nerves after corticosteroid therapy.](image2)