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Optic neuropathy complicating multifocal choroiditis and panuveitis (MFCPU)

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Abstract

Purpose—To describe the clinical characteristics of patients with optic neuropathy complicating multifocal choroiditis and panuveitis (MFCPU).

Design—Retrospective case series.

Methods—8 patients (11 eyes) with MFCPU and optic neuropathy from a single center were reviewed and clinical outcomes described.

Results—The median age of patients was 45 years; 6 patients were women and 7 were white. In the 6 patients with available follow-up, the optic neuropathy was corticosteroid-responsive but required corticosteroid treatment in order to prevent recurrences of optic nerve inflammation and subsequent vision loss. Five patients required immunosuppressive drug therapy during their course. No patients had recurrence of optic neuropathy while receiving immunosuppressive drug therapy. Visual acuity improved or stabilized with treatment in 9 of 11 affected eyes.

Conclusions—Optic neuropathy is an uncommon complication of MFCPU that may result in substantial visual morbidity. Immunosuppressive drug therapy may prevent recurrences of optic neuropathy and subsequent vision loss.

Multifocal choroiditis and panuveitis (MFCPU) is characterized by panuveitis with multiple punched-out lesions at the level of the retinal pigment epithelial.^{1–4} Ocular complications occur commonly in MFCPU and may result in substantial visual morbidity.^{1–5} Optic neuropathy has been reported as a rare complication of MFCPU,⁴ but the characteristics of these patients have not been described previously. We present a series of 8 patients with optic neuropathy complicating MFCPU and describe their course.

Patients with MFCPU and optic neuropathy seen in Ocular Immunology at Wilmer Eye Institute from 1984 through 2005 were studied and clinic characteristics described. The diagnosis of MFCPU was based on the characteristic clinical appearance.^{1,2} Optic neuropathy was diagnosed in eyes with optic disc edema or pallor greater than cupping in patients with an afferent pupillary defect (APD), decreased visual acuity, and either dyschromatopsia or a visual

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field defect consistent with an optic neuropathy. The study was performed with the approval of the Johns Hopkins University Institutional Review Board.

Eight patients with optic neuropathy were identified (Table). Median age at presentation was 45 years. Six patients were women and 7 were Caucasian. Three patients had bilateral optic neuropathy, resulting in 11 affected eyes. All patients had an APD and 7 patients had dyschromatopsia (no data available for Patient 1). Visual field data were available for 7 patients (see Table). Median disease duration was zero months (range: 0 to >10 years). Despite the variable disease duration at presentation to our clinic, all patients had their optic neuropathy diagnosed either at the time of MFCPU diagnosis or early in the disease course. Four patients were receiving prednisone for MFCPU either prior to or at presentation.

Six patients had available follow-up (median follow-up duration = 37 months). All 6 were treated with prednisone 60mg daily with improvement in acuity and color vision, and resolution of optic nerve edema (if present). However, all 6 patients became corticosteroid-dependent and developed recurrences of optic neuropathy when the prednisone was discontinued. In each case of recurrence, treatment was reinstituted with control of the inflammation and at least partial visual recovery. Five patients required immunosuppressive drugs for their optic neuropathy because the prednisone could not be tapered to ≤ 10 mg daily without disease recurrence. None of these patients had recurrences of optic neuropathy after immunosuppressive agents were instituted ($P = 0.02$).

MFCPU is a chronic inflammatory disease that frequently results in ocular complications and loss of visual acuity over time.¹⁻⁵ Although uncommon, we found 8 patients with optic neuropathy complicating MFCPU in our original cohort (12% of patients; 8% of eyes).³ A review of the literature found reports that described optic nerve edema^{2,5} and pallor^{1,4} as complications of MFCPU, but information on other findings consistent optic neuropathy, such as presence of an APD, visual field defects, and dyschromatopsia, were lacking in these reports. It is possible that optic neuropathy is an underreported complication of MFCPU, as to our knowledge, this represents the first series reported in the literature.

During follow-up, the optic neuropathy appeared responsive to but dependent on corticosteroid therapy. Although reinstitution of treatment typically resulted in improvement of the optic neuropathy, the visual recovery after recurrent bouts of inflammation often was not complete. The course of optic neuropathy observed in our patients appears typical to that found in patients with inflammatory optic neuropathy not associated with demyelinating disease.^{6,7} Corticosteroid-dependent optic neuropathy may require immunosuppressive drugs in order to taper the corticosteroids to doses that decrease the likelihood of corticosteroid-related side effects.⁶ The 5 patients that required immunosuppressive drugs had no recurrences of optic nerve inflammation while receiving immunosuppressive drugs, suggesting that this may be the optimal approach to treating these patients.

In summary, optic neuropathy infrequently complicates MFCPU. Our series suggests that this optic neuropathy is steroid-responsive but steroid-dependent, and recurrences of optic nerve inflammation may occur resulting in vision loss. Use of immunosuppressive drugs may reduce recurrences and may help preserve good vision.

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Biography

Jennifer E. Thorne, MD, PhD is assistant professor of Ophthalmology and Epidemiology at the Wilmer Eye Institute, Johns Hopkins University School of Medicine. She completed her ophthalmology residency at Scheie Eye Institute, University of Pennsylvania, her uveitis fellowship at Wilmer, and her PhD in Epidemiology at the Johns Hopkins Bloomberg School of Public Health. Dr. Thorne's research interests include the study of treatment outcomes in inflammatory eye diseases, such as birdshot chorioretinopathy and multifocal choroiditis.

Clinical characteristics of patients with optic neuropathy and multifocal choroiditis with panuveitis (MFPCPU)

Table

Patient Age (y), race, gender	Eye(s)	Initial VA	VA at last follow-up	Presence of APD; Optic disc appearance; Dyschromatopsia; Visual field findings	Disease duration	Presence of ocular inflammation at presentation	Past ocular and medical history	Treatment for optic neuropathy during follow-up	Duration of follow-up (m)
1 46WF	OS	20/15 OD 20/40 OS	20/15 OD 20/40 OS	+APD, optic disc edema & pallor OS; HVF: altitudinal defect OS	14m	0.5+ AC OS; 1+ vitritis OS	CME OS in past; No glaucoma	Prednisone; methotrexate	32
2 41BF	OU*	20/20 OD 20/200 OS	20/20 OD NLP OS	+APD, optic disc edema & pallor; OS; Color: 10/10 OD, 2/10 OS; GVF: cecocentral scotoma, OS	0	None	No glaucoma	Prednisone; cyclophosphamide	171
3 37WF	OD	3/200 OD 20/16 OS	3/200 OD [†] 20/16 OS	+APD, optic disc edema, OD; Color: 1/10 OD, 10/10 OS	0	0.5+ vitritis OD	CNV OD; No glaucoma	No follow-up	0
4 36WF	OD	20/400 OD 20/20 OS	20/400 OD [†] 20/20 OS	+APD, pallor OD; Color: 0/10 OD; 10/10 OS; GVF: central scotoma OD	10m	0.5+ vitritis OU	No glaucoma	No follow-up	0
5 46WF	OS	20/50 OD 20/70 OS	20/100 OD 20/25 OS	+APD, optic disc edema OS; Color: 5/10 OD; 0/10 OS; HVF: central scotoma, MD-22.25 OS	0	0.5+ vitritis OS	CNV OD; No glaucoma	Prednisone; immunran	51
6 53WM	OS	20/60 OD 2/200 OS	20/50 OD HM OS	+APD, pallor OS; Color: 9/10 OD; 0/10 OS; HVF: constriction MD -30.65 OS	0	0.5+ vitritis OU	CME OS in past; No glaucoma OS; elevated IOP OD	Prednisone	8
7 54WF	OU	20/200 OD 20/100 OS	20/125 OD 20/32 OS	+APD, OD, pallor OD>OS; Color: 0/10 OD, 4/10 OS; HVF: constriction OU MD -32.62 OD, -31.38 OS	>7y [‡]	None	ERM OS; Glaucoma OS; Diabetes	Prednisone; mycophenolate mofetil	42
8 58WM	OU	20/200 OD 4/200 OS	20/125 OD 4/200 OS	+APD OD; pallor OU; Color: 6/10 OD, 4/10 OS; GVF: constricted OU; altitudinal OD	>10y [‡]	1+ AC OU; 1+ vitritis OU	Diabetes; Hypertension	Prednisone; mycophenolate mofetil	21

APD = afferent pupillary defect; BF = black female; CME = cystoid macular edema; CNV = choroidal neovascularization; ERM = epiretinal membrane; GVF = Goldman visual field; HM = hand motion; HVF = Humphrey visual field; IOP = intraocular pressure; m = months; MD = mean deviation; NLP = no light perception; OD = right eye; OS = left eye; OU = both eyes; VA = visual acuity; WF = white female; WM = white male; y = years

* At the time of presentation the patient had optic neuropathy OS only. She was lost to follow-up for 7 years. When she returned to our care, she was NLP in the left eye and had optic neuropathy in the right eye (see Case Report 2).

[†]No follow-up was available in these patients so the initial and final visual acuities are the same.

[‡]Review of the patients' records revealed that the optic neuropathy was diagnosed at the same time that the MFPCPU was diagnosed.