between PCV with type 2 CNV and pure fibrinous exudation. Further studies are needed to determine the accurate diagnosis and proper treatment for patients with type 2 CNV associated with PCV.

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Polypoidal choroidal vasculopathy

Polypoidal choroidal vasculopathy

Fumihiko Mori, Shuichiro Eguchi

From the viewpoint of an Asian ophthalmologist

Polypoidal choroidal vasculopathy (PCV) has been recognised for almost two decades. In 1990, Yannuzzi and colleagues first described idiopathic polypoidal choroidal vasculopathy (PCV) macular disorder—a choroidal vasculopathy that causes haemorrhagic and exudative macular degeneration—in 11 patients.¹ The patients had peculiar polypoidal subretinal

vascular lesions associated with serous and haemorrhagic detachments of the retinal pigment epithelium (RPE). In the late 1990s, Yannuzzi and colleagues expanded the clinical spectrum of PCV and established the concept of PCV.^{2,3}

In 1995, Spaide and colleagues identified the precise choroidal abnormalities associated with PCV and examined them using indocyanine green angiography

(IA).⁴ IA showed two basic choroidal vascular changes: a branching network of vessels in the inner choroid and polypoidal vascular dilations at the border of the network of vessels. IA confirmed the definitive diagnosis of PCV. In 1999, using optical coherence tomography (OCT), Iijima and colleagues showed that on images of the inner choroid in eyes with PCV orange-red lesions protruded anteriorly.⁵ These lesions had a sharper peak than serous RPE detachments in the subretina, suggesting that polypoidal vascular lesions in eyes with PCV lie beneath Bruch's membrane and are covered anteriorly by the RPE and Bruch's membrane.⁶ IA and OCT, relatively new diagnostic devices, are useful for understanding the pathophysiology and for diagnosing PCV. However, the cause and the pathogenesis of PCV are not clearly known.

In 1999, Uyama and colleagues described the nature and clinical features of PCV in 32 Japanese patients.⁷ IA showed branching vascular networks with polypoidal dilations at the network terminals beneath the RPE in all patients. These lesions were mostly in the macula (94%), with a few in the peripapillary area. The authors concluded that PCV in Japanese patients differs from that in US patients. In 2002, the same investigators clarified the long-term natural history of PCV.⁸ Fourteen eyes of 12 untreated Japanese patients with PCV were followed prospectively for at least 2 years after the first visit. The authors concluded that PCV is a chronic disease of long duration, and the patients had varying courses. Fifty per cent of the patients had a favourable course; in the other 50%, the disorder persisted for a long time with occasional episodes of bleeding and leakage, resulting in macular degeneration and visual loss. In 2005, the Japanese Study Group of Polypoidal Choroidal Vasculopathy reported the criteria for this disorder in Japanese patients,⁹ based on a fundus examination, IA findings or both. Definitive cases meet at least one of the following criteria: 1) protruding elevated orange-red lesions observed by fundus examination and 2) characteristic polypoidal lesions seen on IA. Probable cases meet at least one of the following criteria: 1) only an abnormal vascular network is seen in IA and 2) recurrent haemorrhagic and/or serous detachments of the RPE are observed.

On fluorescein angiography, eyes with PCV have primarily occult choroidal neovascularisation (CNV) because most components of PCV are beneath the RPE. However, 9% of eyes with PCV in Japanese patients have classic CNV on fluorescein angiography.¹⁰ Otsuji and colleagues evaluated eight eyes of eight Japanese patients with PCV with classic CNV on fluorescein angiography, using IA and OCT.¹¹ All patients had greyish subretinal exudates, which were considered fibrinous. The authors concluded that both type 2 CNV and PCV with fibrin without CNV are present in patients with PCV showing classic CNV. It is important to determine if type 2 CNV is present in eyes with PCV, because the visual prognosis in the presence of type 2 CNV is worse in PCV.

In this issue of the *BJO* (see page 1152), Tamura and colleagues retrospectively reviewed the visual prognosis and ocular

characteristics of 38 eyes with PCV of Japanese patients that appear to have classic CNV on fluorescein angiography.¹² OCT showed subretinal material with moderate reflectivity that corresponded in location to classic CNV in all cases. At the final visit, the subretinal material resolved completely in 14 eyes but resolved incompletely in 24 eyes after photodynamic therapy (PDT). The visual acuity in the group with resolved lesions was significantly better than in the group in which the lesions persisted at the final visit. The authors hypothesised that eyes with persistent lesions have type 2 CNV, and the eyes with resolved lesions have deposition of pure fibrous tissue without CNV. The subretinal material seen before treatment was more frequent in the subfovea in the group with persistent lesions and adjacent to polypoidal lesions in the group with resolved lesions. The authors suggested that although it is difficult to discriminate type 2 CNV from pure fibrin deposition before treatment, type 2 CNV is seen more often in the subfovea and is typically separate from the polypoidal lesions.

Chan and colleagues determined the efficacy of PDT with verteporfin (Visudyne, Novartis, Switzerland) as a treatment for PCV in Asian patients.¹³ Stable or improved vision was achieved in 21 (95%) of the 22 eyes at the 1 year follow-up. Severe visual loss due to massive subretinal haemorrhage occurred in one eye. This report showed that PDT is an effective treatment for PCV. Tamura and colleagues reported that stable or improved vision was achieved in 18 (75%) of the 24 eyes with PCV with type 2 CNV at the final visit. The authors suggested that the treatment effect is limited in eyes with PCV with type 2 CNV.

Recently, the MARINA and ANCHOR studies reported that intravitreal administration of ranibizumab (Lucentis, Genentech, USA), a recombinant, humanised, monoclonal antibody Fab that neutralises all active forms of vascular endothelial growth factor A, prevented vision loss and improved the mean visual acuity with low rates of serious adverse events in patients with CNV secondary to age-related macular degeneration.^{14, 15} The efficacy and safety of this new treatment for PCV should be evaluated. I believe that the diagnosis and treatment of PCV require management by a medical retina specialist using new diagnostic devices

and therapies such as fluorescein angiography, IA, OCT, and PDT.

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