

- 2 Murray DC, Walsh A, Henderson J, Ainsworth JR. Bilateral sixth nerve palsy treated with augmented muscle transposition. *Eye* 2001; **15**: 2001.
- 3 Santiago AP, Rosenbaum AL. Selected transposition procedures. In: Rosenbaum AL, Santiago AP, (eds). *Clinical Strabismus Management*. WB Saunders: Philadelphia, 1999, pp 476–489.
- 4 Rowe FJ. *Clinical Orthoptics*. Blackwell Science: Oxford, 1997.
- 5 Duke Elder W. Recent paralysis of the right superior rectus muscle. In: Duke Elder W (ed). *Textbook of Ophthalmology, Vol. IV, Neurology of Vision — Motor and Optical Anomalies*. Henry Kimpton: London, 1949, pp 4056–4057.
- 6 Helveston EM. Muscle transposition procedures. *Surv Ophthalmol* 1971; **16**: 92–97.

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Sir,

A non-idiopathic case of polypoidal choroidal vasculopathy

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Idiopathic polypoidal choroidal vasculopathy (PCV) is a condition similar to but distinct from age-related macular degeneration. The known underlying ocular predisposing factors for choroidal neovascularisation are absent. Systemic hypertension is a frequent association. We describe a case of PCV where characteristic clinical and angiographic features were precipitated by about of forceful sneezing.

Case report

A 76-year-old Caucasian lady presented to eye casualty complaining of sudden visual distortion in the left eye following a bout of forceful sneezing. She was hypertensive on medication with a blood pressure of 150/86 mmHg at presentation. She had no relevant ocular history. Her best-corrected Snellen visual acuity was 6/9 in each eye. The anterior segments and intraocular pressures were normal. Fundoscopy had revealed some small soft drusen over both maculae. The left had superficial retinal haemorrhages between the disc and the macula and involving the fovea, with surrounding shallow neurosensory retinal detachment. In addition, there was a slightly elevated reddish-orange colour nodule noted at the peripapillary area of the left disc.



Figure 1 Fundus photograph of the left eye 2 weeks after initial presentation, showing the reddish-orange elevated nodule (arrow) in the peripapillary region with superficial retinal haemorrhages extending towards the fovea. Some soft drusen were also found over the macula.

When reviewed in the specialist clinic 2 weeks later, she reported subjective improvement in her vision to 6/6 in each eye. Previously noted neurosensory retinal detachment had resolved with some residual retinal haemorrhages (Figure 1). The differential diagnosis at this stage was age-related peripapillary choroidal neovascularisation. However, the presence of the visible reddish-orange nodule (Figure 1, arrow) strongly suggested PCV.

Fundus fluorescein (Figure 2a and b) and indocyanine green angiography (Figure 3a and b) performed on the same day had confirmed a single hyperfluorescent lesion (arrows) corresponding to the clinically visible reddish-orange nodule. This lesion was well defined with no sign of active leakage as shown from the late phases of both angiograms (Figure 2b and 3b). There was no evidence of age-related choroidal neovascularisation arising from the soft drusen over the macula area. The diagnosis of PCV was therefore made. Laser treatment was not proposed based on clinical and angiographic findings. At 3 months following the initial presentation, the vision continued to improve to 6/5 in the left eye with complete resolution of the retinal haemorrhages but persistence of the reddish-orange nodule.

Comment

Idiopathic PCV was first described in hypertensive black female subjects with characteristic elevated reddish-orange subretinal peripapillary lesions associated with recurrent haemorrhagic retinal pigment epithelial detachments, retinal or vitreous haemorrhages.¹ More recently, case studies have described similar lesions occurring in the macula, the midperipheral, and

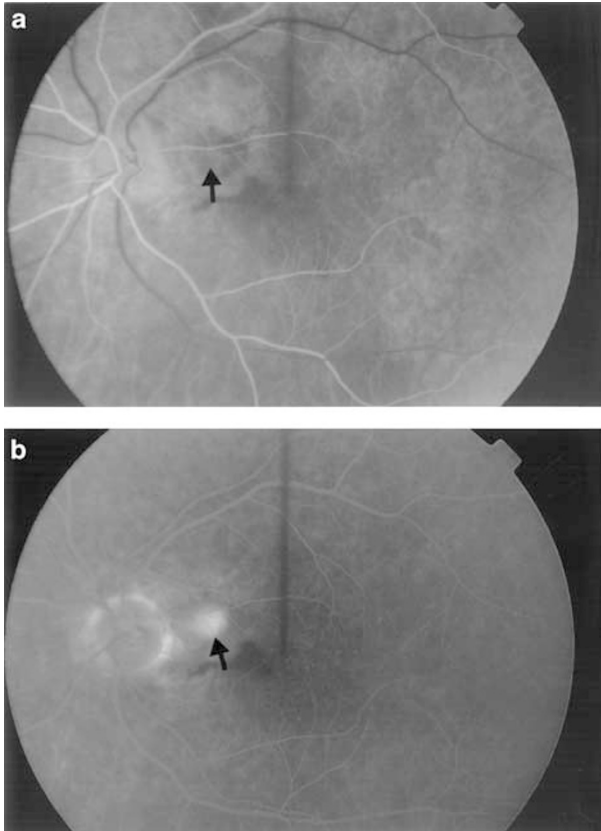


Figure 2 Fundus fluorescein angiogram of the left eye showing a single hyperfluorescent lesion (arrow) corresponding to the reddish-orange nodule and hypofluorescent areas caused by the masking effect of the retinal haemorrhages. The lesion had not demonstrated active leakage as shown in early (a) and late (b) phases of the angiogram.

peripheral retina of Japanese and Caucasians.²⁻⁶ Indocyanine green angiography was useful in defining the deeply located choroidal lesions. Lafaut *et al*⁶ diagnosed 14 of 374 eyes (4%) using indocyanine green angiography, presenting with occult choroidal neovascularisation, as PCV.

The aetiology of idiopathic PCV is unknown. It is believed to be similar to but distinct from age-related macular degeneration.⁵ Progression is slower than age-related macular degeneration and disciform scarring less common.⁵ However, the prognosis remains poor as a result of recurrent haemorrhagic events, which could lead to pigmentary retinal changes or requiring vitrectomy. Conditions that predispose to choroidal neovascularisation such as soft drusen, lacquer cracks, pathological myopia, or intraocular inflammation are conspicuously absent, but coexistence of soft drusen had been reported in more recent literatures.^{3,7} Hypertension is a frequently described systemic association.^{2,3} Other reported systemic associations are diabetes,

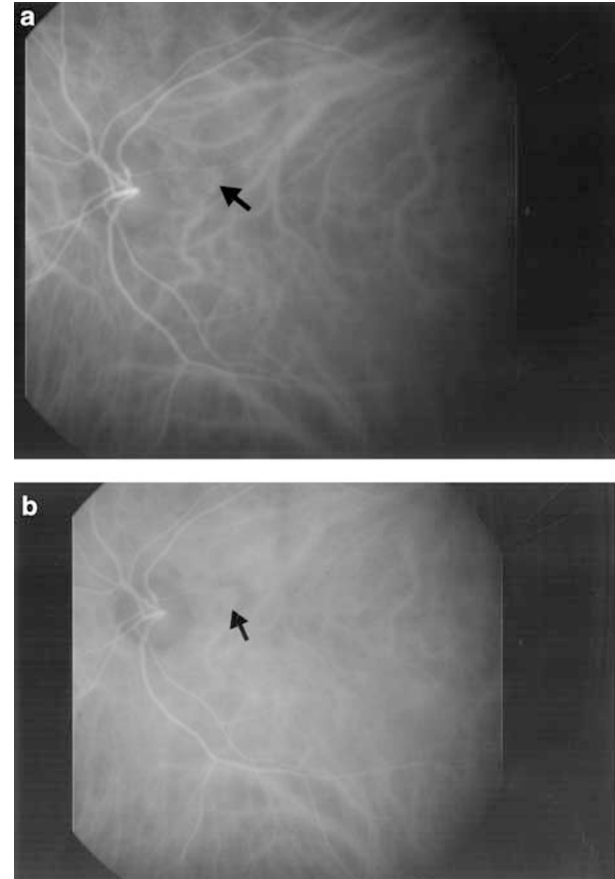


Figure 3 Indocyanine green angiogram of the left eye confirming the presence of a single hyperfluorescent lesion (arrow) of subretinal location. Early (a) and late (b) phases of the angiogram again had not demonstrated active leakage from the lesion.

thrombocytopenia, raised plasma viscosity, and peripheral vasculitis.^{2,3} The occurrence of retinal arterial macroaneurysms and hypertensive retinopathy together with PCV in two elderly black women has been described.⁸ It has been suggested that hypertensive insult to the choroidal vascular bed is a contributory factor in the pathogenesis of PCV.⁸

Histopathological studies on two PCV complexes removed at surgery indicated that they were subretinal choroidal neovascularisation.^{9,10} However, the sharply peaked images obtained on optical coherence tomograms suggest that the lesions lie deep in Bruch's membrane.¹¹ This favours the conventional view that the lesions are most likely large calibre choroidal capillaries with terminal aneurysmal dilatation.³ A recent clinicopathological report of PCV hypothesised that sclerosis of the choroidal vessels could lead to stasis and dilatation of the venules.¹² Subsequent haemorrhage and oedema resulted in the degeneration of the tissue, while the accumulation of blood cells and fibrin might generate

elevation of tissue pressure sufficient to displace the weakened lesion anteriorly.¹²

This case is interesting as the presentation of PCV was precipitated by a bout of forceful sneezing. A broadly similar case was reported by Rosa *et al*¹³ whereby the patient was diagnosed to have PCV, and at a later stage developed massive retinal and subretinal haemorrhage following an episode of severe vomiting. In our case, it is possible that a clinically silent lesion may have been present prior to her sneezing event. She did have risk factors, hypertension, and soft drusen. We postulate that an inner choroidal vascular abnormality possibly associated with hypertensive stress in this patient was further aggravated by the Valsalva effect of sneezing, and may have contributed to the aneurysmal bulging of the choroidal arteries.

References

- 1 Stern RM, Zakov NZ, Zegarra H, Gutman FA. Multiple recurrent serosanguineous retinal pigment epithelial detachments in black women. *Am J Ophthalmol* 1985; **100**: 560–569.
- 2 Lip PL, Hope-Ross M, Gibson JM. Idiopathic polypoidal choroidal vasculopathy: a disease with diverse clinical spectrum and systemic associations. *Eye* 2000; **14**: 696–700.
- 3 Ahuja RM, Stanga PE, Vingerling JR, Reck AC, Bird AC. Polypoidal choroidal vasculopathy in exudative and haemorrhagic pigment epithelial detachments. *Br J Ophthalmol* 2000; **84**: 479–484.
- 4 Uyama M, Matsubara T, Fukushima I, Matsunaga H, Iwashita K, Nagai Y *et al*. Idiopathic polypoidal choroidal vasculopathy in Japanese patients. *Arch Ophthalmol* 1999; **117**: 1035–1042.
- 5 Yannuzzi L, Ciardella A, Spaide RF, Rabb M, Freund KB, Orlock DA. The expanding clinical spectrum of idiopathic polypoidal choroidal vasculopathy. *Arch Ophthalmol* 1997; **115**: 478–485.
- 6 Lafaut BA, Leys AM, Snyers B, Rasquin F, De Laey JJ. Polypoidal choroidal vasculopathy in Caucasians. *Graefes Arch Clin Exp Ophthalmol* 2000; **238**: 752–759.
- 7 Yannuzzi LA, Sorenson J, Spaide RF, Lipson B. Idiopathic polypoidal choroidal vasculopathy. *Retina* 1990; **10**: 1–8.
- 8 Ross RD, Gitter KA, Cohen G, Schomaker KS. Idiopathic polypoidal choroidal vasculopathy associated with retinal arterial macroaneurysm and hypertensive retinopathy. *Retina* 1996; **16**: 105–111.
- 9 MacCumber MW, Dastgheib K, Bressler NM, Chan CC, Harris M, Fine S *et al*. Clinicopathologic correlation of the multiple recurrent serosanguineous retinal pigment epithelial detachments syndrome. *Retina* 1994; **14**: 143–152.
- 10 Terasaki H, Miyake Y, Suzuki T, Nakamura M, Nagasaka T. Polypoidal choroidal vasculopathy treated with macular translocation: clinical pathological correlation. *Br J Ophthalmol* 2002; **86**: 321–327.
- 11 Iijima H, Iida T, Imai M, Gohdo T, Tsukahara S. Optical coherence tomography of orange-red subretinal lesions in eyes with idiopathic polypoidal choroidal vasculopathy. *Am J Ophthalmol* 2000; **129**: 21–26.
- 12 Okubo A, Sameshima M, Uemura A, Kanda S, Ohba N. Clinicopathological correlation of polypoidal choroidal vasculopathy revealed by ultrastructural study. *Br J Ophthalmol* 2002; **86**: 1093–1098.
- 13 Rosa RH, Davis JL, Eifrig CW. Clinicopathologic correlation of idiopathic polypoidal choroidal vasculopathy. *Arch Ophthalmol* 2002; **120**: 502–508.

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Sir,

Spontaneous regression of optic disc neovascularization in Takayasu Arteritis
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A female patient was identified with Takayasu arteritis owing to the presence of disc neovascularization. However, regression of neovascularization may occur without treatment, as in this case, presumably because of vascular remodelling. This phenomenon has not been previously reported.

Case report

A 49-year-old Caucasian female was seen in the ophthalmic department with a 5-year history of diplopia, neck pain, pins and needles in her fingers, and weakness in her arms particularly on lifting. During this time, a vascular surgeon in another centre had discovered right iliac artery disease. An MRI of her carotids and a cervical X-Ray were reported as normal. It was thought that she had severe atherosclerotic disease that was aggravated by her smoking.

She moved house and her new GP discovered that he was unable to find a pulse or blood pressure in either arm. Owing to her varied symptoms, she was referred to the local rheumatology and ophthalmology departments. Carotid, subclavian, and aortic bruits were found. Her arms and right proximal leg pulses were absent. Blood tests showed a normal ESR and haemoglobin, and a raised cholesterol. During her visit to the ophthalmic department, fundal examination revealed neovascularization of her right optic disc and bilateral attenuated arterioles, no other ischaemic signs were seen.