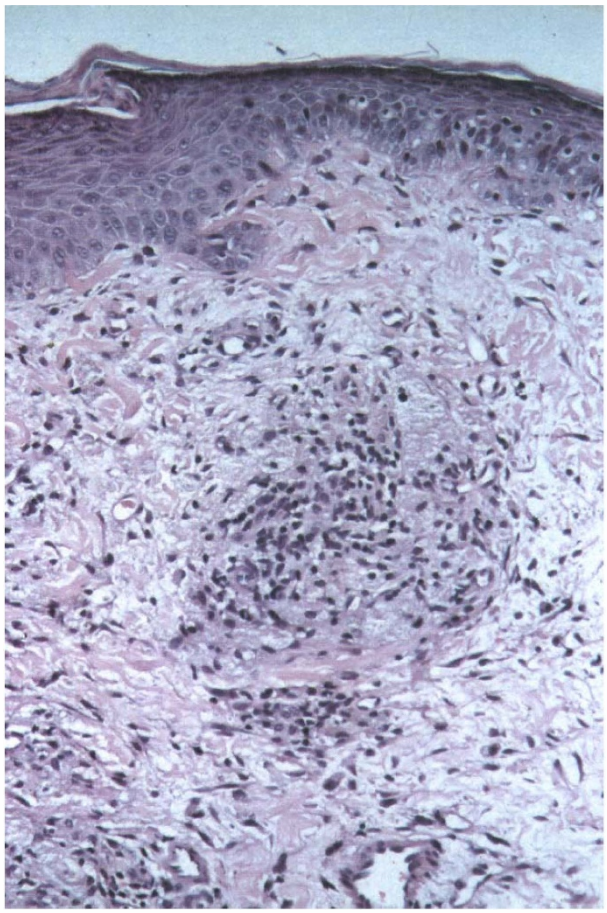

**Fig. 2.** *The post-laser appearances showing a diffuse chronic inflammatory infiltrate within the dermis. While some of the cells present may represent the original large staining cells, these are no longer easily distinguishable within the inflammatory foci. (H&E, original magnification ×200).*

macrophages and the inflammatory response. The patient has been followed-up for 6 months post-treatment, with satisfactory result.

*Comment*

Argon laser photocoagulation has recently been shown to be an effective method of treating xanthelasma lesions without major complications.[5] The $CO_2$ laser has also been used with satisfactory results, although occasional pigmentary changes have been noted. The obvious advantage of the argon laser is its widespread availability and familiarity to ophthalmologists. The advantages over surgery are its use in larger lesions and that the technique is easy and fast to perform.

The histological changes described are consistent with a superficial photocoagulation of the upper skin levels to a depth of 1 mm of dermis, preserving dermal appendages and aiding in the rapid healing of the wound.[4,6] Observation of port-wine stains treated by argon laser have proven such changes to be permanent and stable, suggesting little or no risk of future malignant changes.[6] Xanthelasma is usually an obvious clinical diagnosis, but very rarely other lesions such as xanthogranuloma can simulate the appearances, and if there is any doubt about the nature of the lesion it is better treated with surgical excision. Patients need to be followed-up for up to 6 months to check for immediate recurrence or residual lesion and for the occurrence of scarring. We expect the use of argon laser to treat xanthelasma lesions to become widely accepted in the future.

*References*


1. Bergman R. The pathogenesis and clinical significance of xanthelasma palpebrarum. J Am Acad Dermatol 1994;30:236–42.
2. Arnold HL, Odom RB, James WD. Errors of metabolism. In: Andrews' diseases of the skin, clinical dermatology. 8th ed. Philadelphia: WB Saunders, 1990:602–3.
3. Ullmann Y, Har-Shai Y, Peled IJ. The use of $CO_2$ laser for the treatment of xanthelasma palpebrarum. Ann Plast Surg 1993;31:504–7.
4. Landthaler M, Haina D, Waidelich W, Braun-Falco O. A three-year experience with the argon laser in dermatotherapy. J Dermatol Surg Oncol 1984;10:456–61.
5. Hintschich C. Argonlaserkoagulation von Xanthelasmen. Ophthalmologe 1995;92:858–61.
6. Apfelberg DB, Kosek J, Maser MR, Lash H. Histology of port wine stains following argon laser treatment. Br J Plast Surg 1979;32:232–7.



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Sir,

We welcome the comments made by Mr Adam Booth regarding the family that we recently described.[1] We became aware of the autosomal dominant iridogoniodysgeneses only after submission of our own manuscript, hence their omission from our differential diagnosis. As suggested by Mr Booth, we hope to use the known loci for these conditions as a starting point for our own investigations.

*Reference*


1. Rundle P, Lotery A, Archer DB, McGinnity FG. Familial deafness associated with iris deneration and glaucoma. Eye 1997;11:476–8.



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Sir,

**Central retinal artery occlusion and optic disc drusen**

Central retinal artery occlusion is unusual in young adults.[1] Systemic investigation is indicated to check for migraine, cardiac valvular disease, atrial myxoma, intravenous drug abuse, coagulopathies and collagen

**Table 1.** *Retinal arterial occlusions and optic disc drusen: literature review*

| Author | Age | Sex | Ocular finding |
|---|---|---|---|
| Gifford[19] | 11 | F | Branch retinal arterial occlusion with huge drusen concealing disc |
| Purcell[20] | 50 | M | Sequential bilateral branch retinal artery occlusion; negative systemic investigations |
| Savino[21] | 33 | M | Branch retinal artery occlusion |
| Brown[1] | <30 | F | Branch retinal artery occlusion; taking contraceptive pills |
| Brown[1] | <30 | F | Branch retinal artery occlusion; positive history of migraine; taking contraceptive pills |
| Mustonen[22] | ? | ? | Branch retinal artery occlusion |
| Uehara[23] | 18 | M | Central retinal artery occlusion; rubeosis iridis |
| Newman[24] | 33 | F | Sequential bilateral central retinal artery occlusions 8 years apart; history of migraine |
| Newsom[25] | 48 | M | Sequential bilateral central retinal artery occlusions 5 years apart at high altitude |

vascular diseases. We report on a young man who presented with an acute central retinal artery occlusion from extensive optic disc drusen.

*Case report*

A 39-year-old white man presented with sudden visual loss in the right eye of 1 week's duration. He had a visual acuity of counting fingers at 4 m in the right eye, 6/6 in the left eye, and a moderate right afferent pupillary defect. In the right fundus the retinal arteries were narrowed and the superficial retina was opacified except in the foveola with the resultant cherry-red spot. Optic nerve head drusen were present bilaterally.

The patient was placed on intravenous heparin anticoagulation and the results of extensive systemic investigations were negative (blood sugar, lipids and chemistry; complete blood count; erythrocyte sedimentation rate; activated partial thromboplastin time; prothrombin time; protein C and S; anticardiolipin antibody; haemoglobin electrophoresis; arterial blood gases; antinuclear antibody titre; rheumatoid factor; electrocardiogram; echocardiogram; Doppler carotid ultrasonography). There was no history of migraine, drug abuse, or trauma. Visual status was stable after 4 months of follow-up while maintained on a daily intake of acetyl salicylic acid.

*Discussion*

Optic nerve head drusen occur in 0.3–2% of the general population.[2,3] Drusen represent acellular laminated concretions made of hyaline-like material that is a byproduct of abnormal axonal metabolism.[4] Drusen have predilection for the prelaminar portion of the optic nerve.[4] Most subjects with optic disc drusen are asymptomatic; however, some may have enlargement of the blind spot, nerve fibre bundle defects, and various field defects attributed to 'drusen-associated optic neuropathy'.[5] Drusen of the optic disc are found mainly in the white population.[6] The optic disc area in normal subjects tends to be larger by at least 20% than in subjects with either disc drusen[7] or anterior ischaemic optic neuropathy.[8] Whites have a smaller (by 24%) disc area than blacks or orientals.[9,10]

Anterior ischaemic optic neuropathy is associated with optic disc drusen.[11,12] Both anterior ischaemic optic neuropathy and disc drusen share a similar pathophysiology of axonal crowding from a tight scleral canal. A study in children with optic disc drusen showed that almost half had delayed peripapillary choroidal filling on fluorescein angiography,[13] and this finding is an additional support for the association between disc drusen and ischaemic optic neuropathy.

Disc drusen are also associated with central retinal vein occlusion.[14–18] Six cases of branch retinal artery occlusions have been related to disc drusen[1,19–22] (Table 1) and three cases of central retinal artery occlusion (one associated with migraine and another with high altitude).[23–25] Retinal arterial shunts denoting gradual arterial impedance were reported in 2 cases.[16,17] We propose a dual mechanism for central retinal artery occlusion in the present case: external compression and internal compression. Externally, a small optic disc (more prevalent in whites and especially in discs with drusen) leads to crowding of nerve fibres passing through a tight scleral canal. Internally, the drusen are unyielding material and compress retinal vasculature. Small discs with large drusen predispose to vascular occlusions such as ischaemic optic neuropathy, central retinal vein occlusion and central retinal artery occlusion.

Investigation for optic disc drusen (ultrasonography for buried drusen) needs to be added to the list of investigations performed in young adults with retinal arterial occlusions.

*References*


1. Brown GC, Magargal LE, Shields JA, *et al*. Retinal arterial obstruction in children and young adults. Ophthalmology 1981;88:18–25.
2. Lorentzen SE. Drusen of the optic disc: a clinical and genetic study. Acta Ophthalmol (Copenh) 1966;90(Suppl):1–179.
3. Rosenberg MA, Savino PJ, Glaser JS. A clinical analysis of pseudopapilledema. I. Population, laterality, acuity, refractive error, ophthalmoscopic characteristics, and coincident disease. Arch Ophthalmol 1979;97:65–70.
4. Tso MOM. Pathology and pathogenesis of drusen of the optic nerve head. Ophthalmology 1981;88:1066–79.
5. Scholl GB, Song H-S, Winkler DE, Wray SH. The pattern visual evoked potential and pattern electroretinogram in drusen-associated optic neuropathy. Arch Ophthalmol 1992;110:75–81.
6. Mansour AM, Hamed LM. Racial variation of optic nerve diseases. Neuro-ophthalmology 1991;11:319–23.
7. Mullie MA, Sanders MD. Scleral canal size and optic nerve head drusen. Am J Ophthalmol 1985;99:356–9.
8. Mansour AM, Shoch D, Logani S. Optic disk size in ischemic optic neuropathy. Am J Ophthalmol 1988;106:587–9.

9. Mansour AM. Racial variation of optic disc size. Ophthalmic Res 1991;23:67–71.
10. Mansour AM. Racial variation of optic disc parameters in children. Ophthalmic Surg 1992;23:469–71.
11. Newman WD, Dorrell ED. Anterior ischemic optic neuropathy associated with disc drusen. J Neuro-ophthalmol 1996;16:7–8.
12. Gittinger JW, Lessell S, Bondar RL. Ischemic optic neuropathy associated with optic disk drusen. J Clin Neuro-ophthalmol 1984;4:79–84.
13. Erkkila H. The central vascular pattern of the eyeground in children with drusen of the optic disk. Graefes Arch Klin Exp Ophthalmol 1976;199:1–10.
14. Green WR, Chan CC, Hutchins GM, Terry JM. Central retinal vein occlusion: a prospective histopathologic study of 29 eyes in 28 cases. Retina 1981;1:27–55.
15. Austin JK. Optic disk drusen and associated venous stasis retinopathy. J Am Optom Assoc 1995;66:91–5.
16. Chern S, Magargal LE, Annesley WH. Central retinal vein occlusion associated with drusen of the optic disc. Ann Ophthalmol 1991;23:66–9.
17. Borruat FX, Sanders MD. Vascular anomalies and complications of optic nerve drusen. Klin Monatsbl Augenheilkd 1996;208:294–6.
18. Karel I, Otradovec J, Peleska M. Fluorescence angiography in circulatory disturbances in drusen of the optic disk. Ophthalmologica 1972;164:449–62.
19. Gifford H. An unusual case of hyaline bodies in the optic nerve. Arch Ophthalmol 1895:24:395–401.
20. Purcell JJ, Goldberg RE. Hyaline bodies of the optic papilla and bilateral acute vascular occlusions. Ann Ophthalmol 1974;6:1069–76.
21. Savino PJ, Glaser JS, Rosenberg MA. A clinical analysis of pseudopapilledema. II. Visual field defects. Arch Ophthalmol 1979;97:71–5.
22. Mustonen E. Pseudopapilledema with and without verified optic disc drusen: a clinical analysis. II. Visual fields. Acta Ophthalmol (Copenh) 1983;61:1057–66.
23. Uehara M, Inomata H, Yamama Y, *et al*. Optic disc drusen with central retinal artery occlusion. Jpn J Ophthalmol 1982;26:10–7.
24. Newman NJ, Lessell S, Brandt EM. Bilateral central retinal artery occlusions, disk drusen, and migraine. Am J Ophthalmol 1989;107:236–40.
25. Newsom RSB, Trew DR, Leonard TJK. Bilateral buried optic nerve drusen presenting with central retinal artery occlusion at high altitude. Eye 1995;9:806–8.
26. Auw-Haedrich C, Mathieu M, Hansen LL. Complete circumvention of central retinal artery and venous cilioretinal shunts in optic disc drusen. Arch Ophthalmol 1996;114:1285–7.

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Sir,

**Peripapillary lesions causing blind spot enlargement in a case of multiple evanescent white dot syndrome**

Multiple evanescent white dot syndrome (MEWDS) is a multifocal chorioretinal inflammatory disease that was first reported by Jampol *et al*. in 1984.[1] Their original case series described 11 young adults with a unilateral decrease in visual acuity. Ophthalmoscopic findings revealed white dots distributed at the level of deep retina and/or retinal pigment epithelial layer (RPE) – the site thought to be primarily affected in MEWDS. In addition there was macular granularity.

Since the original description many cases of MEWDS have been described[2–5] and the clinical picture better defined. Most cases are preceded by a viral illness, and symptoms of photopsia are common.[2] Vitritis, optic neuritis and enlargement of the physiological blind spot are also typical features. While the exact aetiology of MEWDS remains unknown, the cause of the enlarged blind spot has been the focus of several studies. Takeda *et al*.[6] suggested optic nerve dysfunction as the cause. Fletcher *et al*.[7] maintained that peripapillary retinal dysfunction was the mechanism after studying patients with blind spot enlargement and normal optic discs. The following case of MEWDS is unusual in that peripapillary lesions were demonstrable (and thought to be responsible for the enlarged blind spot) in addition to an undoubted optic neuritis.

*Case report*

A 34-year-old woman presented with a 2 week history of photopsia in the left visual field that was associated with reduced visual acuity and floaters in the left eye. Visual acuity in the right eye was 6/4 and in the left 6/36. A left relative afferent pupillary defect, red desaturation and blind spot enlargement on automated perimetry was present. Fundal examination revealed a vitritis, papillitis, and marked peripapillary nerve fibre layer swelling on the left, as well as multiple mid-peripheral white lesions at the level of the retinal pigment epithelium (RPE) (Fig. 1). Fundal fluorescein angiography (FFA) in the acute phase showed early hyperfluorescence and late staining of lesions typical of MEWDS. There was also extensive leakage of dye from the optic nerve head consistent with a papillitis, and no peripapillary lesions could be recognised (Fig. 2).

The patient was commenced on prednisolone 80 mg daily and 1 week later her visual acuity improved to 6/18. The left relative afferent defect was less obvious as were the chorioretinal lesions on fundoscopy. The

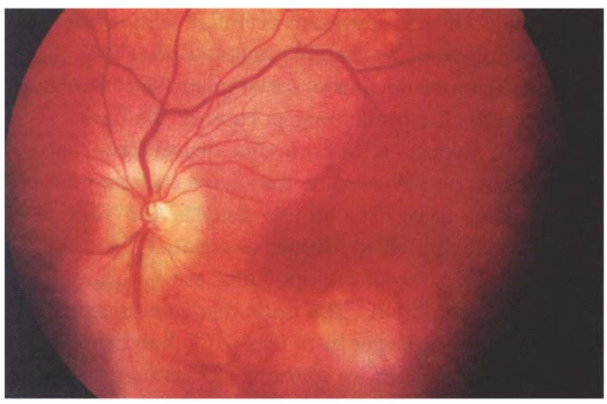

**Fig. 1.** *A fundus photograph of the left eye at presentation, showing swelling of the optic disc, and peripapillary nerve fibre swelling. Multiple mid-peripheral whitish lesions, best seen supero-temporally, are also present. (The circular pale area infero-temporal to the macula is artefactual.)*