'Prion disease'² refers to progressive, subacute, degenerative disease of the CNS, and in addition to CJD includes Gerstmann–Straussler–Scheinker disease (GSS), kuru, fatal familial insomnia and familial atypical dementia. Similar diseases which occur in animals include scrapie in sheep and bovine spongiform encephalopathy (BSE) in cattle. What prion diseases have in common is an abnormal protease-resistant isoform of the prion protein (PrP) which is normally expressed by neurons. Genetic analysis of the PrP gene can help to differentiate familial and sporadic disease. In addition, the homozygous methionine genotype at codon 129 of the PrP gene occurs in all cases of variant CJD but only 70% of sporadic cases, and further work is being undertaken to look at the susceptibility to variant CJD in individuals who are heterozygous or valine homozygous at codon 129. However, to date definitive diagnosis of CJD can only be made histologically. PrP glycotyping on western blot may help to differentiate variant and sporadic disease.^{2,17,18}

With improved awareness of CJD and its clinical manifestations, ophthalmic signs may prove a useful aid in the early diagnosis of this condition.

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A.M. McElvanney
 Eye Department
 Royal Surrey County Hospital
 Guildford
 Surrey GU2 5XX, UK
 M. G. Boodhoo
 Eye Department
 Saint Peter’s Hospital
 Chertsey
 Surrey KT16 0PZ, UK
 Miss A.M. McElvanney ✉
 Eye Department
 St George’s Hospital
 Blackshaw Road
 London SW17 0QT, UK

Sir,

Indocyanine green angiographic imaging of central retinal artery occlusion with cilioretinal sparing
 Central retinal artery occlusion (CRAO) is an ophthalmic emergency that generally results in a poor prognosis. Vision is severely affected unless a cilioretinal artery is present to supply circulation to the macular area. Although fundus fluorescein angiography (FFA) clearly demonstrates retinal vascular defects associated with CRAO, indocyanine green (ICG) angiographic features of this clinical entity are not well documented. Here we describe the ICG angiographic images and the choroidal and retinal alterations in a case of CRAO with cilioretinal sparing.

Case report

A 66-year-old woman with a 5-year history of hypertension reported the sudden onset of painless decreased vision in the right eye, which had started 12 h earlier. On examination her best corrected visual acuity was counting fingers with her right eye and 20/20 in the left eye. A right, relative afferent pupillary defect was present. Anterior segment examinations were normal and intraocular pressure (IOP) was 14 mmHg in both eyes. Fundus examination of the right eye showed segmentation of the blood column in the retinal arteries and diffuse whitening of the retina, except for the localised area between the fovea and the optic disc, indicating CRAO with cilioretinal sparing. Examination

Fig. 1. Fundus fluorescein angiography demonstrating the filling of the cilioretinal artery and the lack of filling of the retinal arteries.

of the left fundus revealed mild hypertensive retinopathy. Immediate, anterior chamber paracentesis and ocular massage were carried out. After thorough systemic examination the patient was found to have uncontrolled systemic arterial hypertension. All other test results were negative. The cause of the CRAO was thought to be systemic hypertension and therefore systemic treatment with nifedipine 30 mg daily and aspirin 300 mg daily was initiated. At the 13th hour of occlusion both FFA and ICG angiography were performed using a Topcon IMAGENET H 1024 Digital Imaging System (Topcon, Tokyo, Japan).

Both the filling of the cilioretinal artery and the lack of filling of the retinal arteries were observed on FFA (Fig. 1). The ICG angiography findings were studied in three phases. The early-phase ICG angiogram demonstrated normal background fluorescence only in the region of the cilioretinal artery, which contrasted sharply with the adjacent hypofluorescent areas supplied by the central retinal artery (CRA) (Fig. 2a). Retinal circulation was virtually non-existent in the remainder of the fundus apart from the flow in the cilioretinal artery. The fluorescence of the branch of the inferior temporal vein draining the perfused area was noted before any dye appeared in the retinal arteries. The left eye demonstrated normal choroidal fluorescence and filling pattern in the retinal and choroidal vessels. In the middle phase of the ICG angiogram, the retinal vessels of the right eye remained poorly filled at nearly 8 min after injection. The area supplied by the cilioretinal artery was normal, while the surrounding area supplied by the CRA was hypofluorescent. ICG angiography of the left eye was normal (Fig. 2b). The late-phase ICG angiogram of the left eye showed a silhouette of retinal vessels that no longer contained dye, against a background of choroidal fluorescence. In stark contrast to this, the right eye showed staining of the walls of the retinal arteries and veins, even at 45 min after injection, a finding which indicated vascular damage (Fig. 2c).

(a)

(b)

(c)

Fig. 2. (a) Early-phase indocyanine green (ICG) angiogram demonstrating normal background fluorescence only in the territory of the cilioretinal artery. The retinal circulation is virtually non-existent except for the cilioretinal artery, and fluorescence of the branch of the inferior temporal vein draining the perfused area is noted before any dye appears in the retinal arteries. (b) Middle phase of the ICG angiogram demonstrating that the retinal vessels are still poorly filled. The area supplied by the cilioretinal artery was normal while the surrounding area was hypofluorescent. (c) Late-phase ICG angiogram demonstrating staining of the wall of the retinal arteries and veins even at 45 min in the right eye.

Although ICG angiographic studies of CRAO have been published,^{1,2} CRAO with cilioretinal artery sparing has not been reported to date. Cilioretinal arteries are usually derived from short posterior ciliary arteries but have also been shown to arise directly from the choroidal vessels in the subhuman primate model.³ Approximately 25% of eyes with CRAO have cilioretinal sparing of part of the macula.⁴

In our case, ICG angiography allowed us to follow all the stages of CRAO with cilioretinal sparing. The early-phase ICG angiogram showed normal background fluorescence only in the region of the cilioretinal artery, which contrasted sharply with the adjacent hypofluorescent areas supplied by the CRA. Although vascular filling defects and blocking substances such as pigment, haemorrhage and exudation have been included in the causes of hypofluorescence seen in ICG angiograms,⁵ special attention to CRAO with cilioretinal sparing has not been reported. Diffuse pallor of the retina due to intracellular oedema, cellular necrosis and accumulation of cellular debris may cause hypofluorescence surrounding the area of cilioretinal sparing in which normal background fluorescence is present. The retinal arteries did not fill completely, even in the middle phase, but our most striking finding was the persistent staining of the retinal vessels at 45 min after injection. In contrast, FFA showed no fluorescence at the late stage. This persistent staining may be caused by a combination of the protein-binding properties of ICG and the retinal vessel damage, which can not be documented with FFA. ICG angiography does not reveal the details provided by FFA when imaging the normal retinal vasculature, but our case shows that, with some pathological conditions, ICG angiography may highlight abnormalities of the retinal and choroidal circulation.

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Gürsel Yılmaz
Dilek Dursun
Pinar Aydın
Baskent University, School of Medicine
Department of Ophthalmology
Fevzi Çakmak Cad. 10. Sok. No:45
06490 Bahçelievler
Ankara, Turkey

Dr Gürsel Yılmaz ✉
Mesrutiyet Cad. 44/15
06420 Kizilay
Ankara, Turkey
Tel: +90 312 4359408
Fax: +90 312 2237333
e-mail: dyilmaz@dialup.ankara.edu.tr

Sir,

Bilateral choroidal metastases as the first sign of metastatic gestational choriocarcinoma

The uvea is the most common site of ocular metastases, with the choroid accounting for approximately 80% of all metastatic ocular disease.¹ Uveal metastases represent the most common uveal malignancy, being more common than primary uveal melanoma.^{2,3} Uveal metastases are spread haematogenously, with the posterior pole (particularly the submacular choroid) most often involved.⁴ A metastatic tumour of the uvea is the initial clinical manifestation of systemic cancer in approximately 17% of patients.⁵

Gestational trophoblastic neoplasia is the term applied to choriocarcinoma and related tumours. Choriocarcinoma develops in 1 in 40 000 term pregnancies; in contrast, it develops in approximately 3–5% of patients with a molar pregnancy. In approximately 50% of patients with choriocarcinoma the disease develops following a hydatidiform mole; in approximately 25%, following a term pregnancy; and in 25%, following an abortion (spontaneous, therapeutic or ectopic). Aside from direct tissue examination, one of the hallmarks of diagnosis is an elevated urine or serum titre of human chorionic gonadotrophin (hCG), the 'pregnancy hormone', which is produced by this tumour in large quantities. Non-gestational choriocarcinomas derive from germ cells of the testis or ovary. Because of a propensity for haematogenous spread, all the blood-rich organs are at significant risk for metastatic disease. Metastatic disease can be found in the lung, genitourinary system, brain, liver, spleen and gastrointestinal tract.⁶

Gestational choriocarcinoma rarely metastasises to the eye, and only few cases of metastasis to the choroid have been reported.^{7–14} We report here an unusual case of gestational choriocarcinoma that presented as bilateral choroidal metastases and serous retinal detachment.

Case report

A 41-year-old woman presented to the retina service, King Abdulaziz University Hospital, with a 2-month history of decreased vision in both eyes. On examination, the patient was found to have a visual acuity of counting fingers in both eyes. Slit-lamp examination yielded normal results and the intraocular pressure was 12 mmHg bilaterally. Dilated fundoscopy showed bilateral, multiple, yellow, elevated choroidal lesions involving the macular and juxtapapillary areas with total bullous sensory retinal detachments (Fig. 1). The choroidal lesions showed moderate- to high-amplitude internal reflectivity on ultrasonography. A fluorescein angiogram