LETTERS TO THE EDITOR

Benign fleck retina

EDITORS,—We report a case of flecked retinal dystrophy in a child of mixed Australian abo-
riginal and white descent. Both fundi showed widespread discrete yellow-white fleck lesions
at the level of retinal pigment epithelium, extending to the far periphery but sparing the
macular region. Visual acuity was normal and the electroretinogram showed no abnormal-
ity. To our knowledge no other family members are affected, and there is no history of
consanguinity between the parents. The phenotype appears similar to the 'benign familial
fleck retina' described by Aish and Dajani,1 and we believe this to be the first published
report of such a case since the original description in 1980.

CASE REPORT

A 12-year-old girl was referred after her
optometrist noted areas of retinal abnormality
on ophthalmoscopy after a routine refraction.
She complained of occasional headaches, but
had no specific ocular symptoms and was in
good general health. Drug or substance abuse
was denied. There was no history of maternal
illness or nutritional deficiency during preg-
nancy, and the patient was born at full term.
Visual and general development were report-
edly normal during infancy and early child-
hood, and the mother noted no signs of poor
daytime or night vision in any of her three
children.

The child was of mixed ethnic origin, the
non-consanguineous parents both being of
mixed Australian aboriginal and white
descent, and there was no family history of
ocular or systemic disease other than the
recent development of maturity onset dia-
betes mellitus in the mother.

General examination found no evidence of
skin depigmentation or vitiligo. Visual acuities
were 6/6 right with +0.50 DS/-1.00
DC×180 degrees, 6/6 left with +0.50 DS.
Slit-lamp examination of anterior segments
and vitreous was normal. Funduscopy
revealed a striking pattern of multiple yellow-
white flecks situated at a level deep to the
retinal vessels, affecting both fundi in a
symmetrical pattern (Fig 1). The lesions were
distributed in a concentric pattern around the
posterior fundus but sparing the optic disc,
papillomacular bundle, and the peripapillary
area for a distance of 1 disc diameter except
inferiorly, where there was involvement to the
margin of the optic disc. No part of the equa-
torial, peripheral, or far peripheral retina
was spared. The shape of the flecks was highly
variable. The most centrally located lesions
were roughly round or oblong in shape,
and approximately the diameter of a third order
arteriole. With increasing distance from the
central macula, the flecks tended to become
much larger and confluent, up to 1/2 disc
diameter in size across the largest dimension.
The more peripheral flecks showed greater
variability in shape, with linear, geographical,
or amoeboid outlines and smooth sinuous
margins. On stereoscopic slit-lamp biomicro-
scopy, the fleck lesions appeared flat, with
no evidence of accumulated material within
or anterior to retinal pigment epithelial cells.
There was no evidence of pigment migration,
although several of the larger lesions enclosed
areas of normally pigmented retina, giving the
appearance of a central pigment clump (Fig
1C, arrowed). No calcification was seen, and
choroidal vessels were not visible at the base
of the flecks. Fundus fluorescein angiography
revealed patchy, fine, irregular hyperfluores-
cence throughout the fundus which did not
 correspond to the fleck lesions (Fig 2).

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neither a 'window defect' suggesting depigmentation, nor hypofuorescence, which might suggest an abnormal accumulation of material within retinal pigment epithelial cells. Instead, the mild generalised irregular hypofuorescence suggests merely a diffuse abnormality of retinal pigment epithelium.

The occurrence of a marbelised fundus in asymptomatic patients is rare. Aish and Dajani have described an Arab Palestinian family with clinical features which appear to closely resemble those of our patient. In this pedigree, the parents were phenotypically normal first cousins. Seven out of 10 of their offspring showed massive invasion of both fundi by bright white or yellow flecks lesions situated behind the retinal blood vessels, and always sparing the macula. Visual findings were normal in all cases. The probable mode of inheritance within this family was autosomal recessive, since both sexes were involved, and the consanguineous parents were unaffected. Krogh et al have described an asymptomatic 31-year-old woman with normal visual acuity, with bilateral retinal flecks in the mid periphery of both eyes. The flecks became more dense in the periphery, where they formed a palisade pattern quite unlike that of our case. Functional testing revealed an absent EOG light rise in one eye but was otherwise normal. More recently, a case of bilateral 'breadcrumb' flecked retinopathy with normal fluorescein angiography and normal electrophysiological findings has been reported in a 9-year-old girl. However, this child also had an idiopathic seizure disorder which had been controlled medically for 6 years, subnormal intelligence, gross motor and developmental delay, and esotropia. The size and shape of the retinal flecks in this case are not described in detail, but the published photographs appear to demonstrate a more uniform size and more irregular margins to the flecks than in our patient, with a more linear distribution of flecks and a greater area of normal appearing retina between the flecks.

A marbelised fundus appearance has also been reported as a rare finding in Leber's congenital amaurosis. In this variant, yellowish lesions are seen deep to the retinal vessels in a perieriaterial distribution, and there may be associated systemic abnormalities, including medullary cystic renal disease (juvenile nephropathies). The other clinical features and absent ERG response of Leber's amaurosis make confusion with our case unlikely. However, it is interesting to note in such cases that a marbelised fundus may be incidental to visual functional abnormalities.

We suggest that our case represents either a new mutation of the condition described, or possibly an autosomal recessive disorder, since both parents are phenotypically normal.

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Brown's syndrome as a complication of cardiopulmonary resuscitation

EDITOR—Brown's syndrome is a well recognised ocular motility disorder which may be congenital or acquired. Regardless of aetiology it manifests itself clinically with restriction to both active and passive elevation in adduction, minimal or only slight limitation to elevation in abduction, occasionally a downshoot of the affected eye in adduction and, in more severely affected cases, a primary position hypotropia with an associated abnormal head posture. The head posture consists of a chin-up head position with a face turned away from the affected side or a variable head tilt. Other features less commonly seen are a 'V' pattern resulting from divergence in upgaze and widening of the palpebral fissure on adduction.1

CASE REPORT

We report a case of acquired Brown's syndrome in a 2-year-old girl without a history of ocular or head trauma. She accidentally fell into the family swimming pool and was found cyanotic, face down in the water. Her mother, trained in cardiopulmonary resuscitation, rescued her from the pool and subsequently recognised her using nasal compression, mouth to mouth ventilation, and cardiac massage. The patient was transferred to the Children's Hospital of Pittsburgh and, after a short period of artificial ventilation, made a full recovery. A computed tomography brain scan showed no abnormalities.

After hospitalisation it was noted that the child had developed a mild chin-up head posture. One week later her vision was 20/30 in each eye using Allen figures, she was orthophoric in the primary position of gaze, had a chin-up head posture without head tilt or face turn, and ocular versions revealed limitation of elevation in adduction (Fig 1).

There was no evidence of superior oblique muscle overaction or downshoot in adduction. In addition, the right superior rectus muscle did not accommodate, and there was some divergence in upgaze which helped to differentiate this entity from an isolated left inferior oblique paresis. She demonstrated 100 seconds of arc stereocuity and had normal fusion for both distance and near using the Worth 4 dot test in the primary position of gaze. Magnetic resonance imaging of the orbits was normal and did not reveal any evidence of trochlear disinsertion or swelling. The orbital floor was intact. When the patient was considered sufficiently mature we performed forced duction testing under local anaesthesia which confirmed the diagnosis. The patient was 3 years old at this time. We did not feel it necessary to subject the patient to general anaesthesia when she first presented in order to confirm the diagnosis, particularly in light of her near drowning event.

This patient has been followed for 18 months, and the restriction in elevation in adduction has improved significantly. As the patient did not have a significant head tilt and was orthophoric in primary position surgical intervention was not required.

COMMENT

Acquired Brown's syndrome has been reported following traumatic events occurring in the region of the trochlea; these include peribulbar anesthesia,2 orbital surgery,3 orbital roof fracture with superior oblique

Figure 3 Full field Ganzfeld electroretinogram (ERG). Upper traces show scotopic ERG elicited by a single white flash in the dark adapted eye: A- wave latency 25-5 ms RE, 25-7 ms LE (normal range 24-4-28-1 ms), amplitude 121-0 µV RE, 131-0 µV LE (normal range 56-1-185-2 µV). B- wave latency 29-9 ms RE, 47-8 ms LE (normal range 45-6-53-9 µV) and amplitude 549-0 µV RE, 616-0 µV LE (349-0-676-9 µV). Lower traces show photopic ERG response to a single white flash: A- wave latency 16-6 ms in each eye (normal range 16-5-18-9 ms) and amplitude 27-0 µV RE, 28-0 µV LE. B- wave latency 30-7 ms RE, 31-0 ms LE (27-5-31-8 ms) and amplitude 121-0 µV RE, 151-0 µV LE (39-1-207-1 µV). All normal range values represent mean (2 SD).