

Rubella maculopathy

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SUMMARY Three patients with known history of congenital rubella and sudden decrease of vision are presented. Two of the 3 patients had previous eye examinations which showed typical rubella (salt and pepper) retinopathy. All 3 showed macular lesions associated with presumed subretinal neovascularisation.

The ocular and systemic manifestations of congenital rubella were first recognised by Gregg in 1941.¹ His description included the triad of cataracts, congenital heart disease, and deafness of infants born in an Australian epidemic of rubella. However, no mention of fundus changes was made. Gregg attributed the first fundus observation to Mitchell,¹ who in 1939 described a child with left monocular cataract. Mitchell stated that the fundus of the patient's right eye appeared pale, and that some scattered irregular spots of pigment were observed. Previous authors have stressed that visual function remains stationary and that rubella retinopathy is a benign and nonprogressive condition.^{2–6} The purpose of this report is to demonstrate that this previous concept is not necessarily valid in all instances.

Case reports

CASE 1

The first case is that of an 11-year-old white male with a prenatal history of a maternal exposure to rubella during the first trimester. The length of pregnancy and delivery was normal. The patient had no significant systemic disease, but was diagnosed as having profound deafness at 5 weeks of age. There was no history of ocular trauma. The patient's previous ocular record shows that in 1975 the visual acuity was 20/20 in each eye. In February 1976 the vision in the right eye was corrected to 20/20 while the best correction in the left eye was 3/400. External examination was unremarkable except for some early bilateral band keratopathy. The anterior segment showed some scattering of

pigment on the anterior lens capsule in the left eye and a snowflake cataract in the right eye. Ophthalmoscopic examination of the right macula showed some pigment mottling with loss of the foveolar reflex (Fig. 1a).

In the left macula there was a yellowish fibrotic scar with a small punctate haemorrhage beneath the sensory retina (Fig. 1b). In the early laminar phase of the fluorescein angiogram diffuse and small pigment epithelial window defects in the macular region can be seen (Fig. 1c). Late phase of the angiogram shows increased hyperfluorescence due to the staining of the scar tissue, with some blockage of the background fluorescence by punctate haemorrhage beneath the sensory retina (Fig. 1d).

CASE 2

The second case is that of an 11-year-old black male with a prenatal history of maternal rubella during the first trimester of pregnancy. At 18 days of age the child was admitted to hospital for surgical repair of a patent ductus arteriosus and at that time was found to have a severe hearing defect. In March 1976 the patient complained of decreased vision in the left eye. There was no history of ocular trauma. Vision in the right eye was 20/20, while vision in the left eye was correctable to only 20/400. Anterior segments of both eyes were normal.

Ophthalmoscopic examination of the left posterior pole showed diffuse pigment epithelial mottling with a salt and pepper appearance. In the macular region there was a yellowish fibrotic scar (Fig. 2a). The fluorescein angiogram of the left posterior pole showed both hyperfluorescence and blocked fluorescence due to the diffuse pigment epithelial mottling. There was an increased hyperfluorescence directly within the foveolar region due to staining of the disciform scar (Fig. 2b).

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Fig. 1a Right macula: pigment mottling with loss of the foveolar reflex.



Fig. 1b Left macula: a yellowish fibrotic scar with a small punctate haemorrhage beneath the sensory retina.

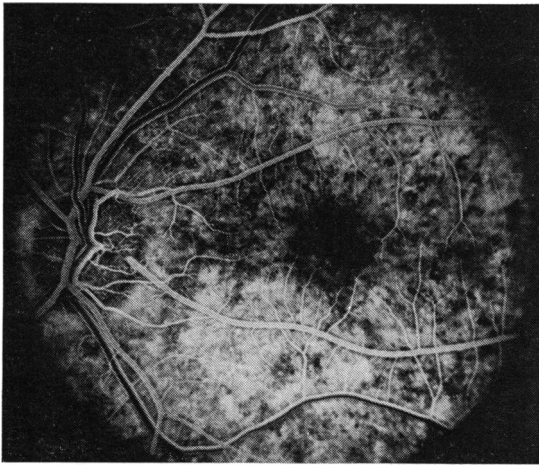


Fig. 1c Early laminar phase of the fluorescein angiogram reveals diffuse small pigment epithelial window defect. There is increased blockage in the macular region secondary to the fibrotic tissue.



Fig. 1d Late phase of the fluorescein angiogram shows increased hyperfluorescence due to staining of the scar tissue with blockage of the background fluorescence by the punctate subretinal haemorrhage.

CASE 3

Case 3 is that of a 12-year-old white male with a prenatal history of maternal exposure to rubella virus during the first trimester of pregnancy. The patient was found to be deaf at 4 months of age. In 1976 the best corrected visual acuity was reported as 20/30 in the right eye and 20/20 in the left eye.

In November 1977 the patient's best corrected visual acuity in the right eye was found to be 20/400,

while vision in the left eye was 20/20. There was no history of ocular trauma. The external examination as well as the anterior segment examination were within normal limits.

The midperiphery of the right eye showed a typical salt and pepper pigmentary mottling (Fig. 3a). The left macula showed generalised mottling of the pigment epithelium with loss of both the macular ring and foveolar reflexes⁷ (Fig. 3b).



Fig. 2a *Left macular region: diffuse pigment epithelial mottling with a salt and pepper appearance and a yellowish fibrotic disciform scar.*

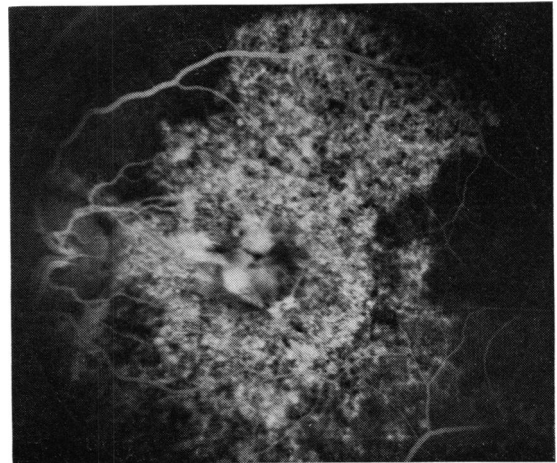


Fig. 2b *Venous phase of the fluorescein angiogram reveals both hypo- and hyperfluorescence due to the pigment epithelial mottling. Increased hyperfluorescence is seen within the foveolar region due to staining of the disciform scar.*

In the right posterior pole there was a small serous detachment of the sensory retina with a small subretinal haemorrhage. A greyish discoloration could be seen inferior to the haemorrhage (Fig. 3c).

Early in the laminar phase of the angiogram

there was an increased blockage of fluorescence in the macula due to subsensory retinal and subretinal pigment epithelial haemorrhage (Fig. 3d). In the venous phase of the angiogram there was still blockage of the background fluorescence. Also seen was an area of increased hyperfluorescence due



Fig. 3a *Midperiphery of the right eye: a typical salt and pepper pigmentary mottling.*



Fig. 3b *Left macula: generalised mottling of the pigment epithelium with loss of both the macular ring and foveolar reflexes.*



Fig. 3c *Right macular region: a small serous detachment of the sensory retina with subretinal haemorrhage. A greyish discoloration can be seen inferior to the haemorrhage.*

to fluorescein leakage, presumably from subretinal neovascularisation (Fig. 3e).

Discussion

These 3 cases of rubella maculopathy, in addition to the 3 recently reported by Deutman and Grizzard,⁸ point out that disciform formation secondary to subretinal neovascularisation is a definite complication which can develop in patients with rubella retinopathy. Rubella maculopathy must now be added to the ever expanding list of disorders associated with subretinal or subretinal pigment epithelial neovascularisation. At present we have no histopathological evidence that these disciform scars in patients with rubella maculopathy are due to the persistence of the rubella virus. The authors feel that the pre-existing disease at the level of the retinal pigment epithelium and Bruch's membrane is most likely the cause for the development of subretinal neovascularisation and the eventual disciform scar.

In conclusion, we have studied 3 cases of congenital rubella retinopathy with macular lesions, associated with presumed subretinal neovascularisation. Although a typical subretinal neovascular membrane was not seen in our cases or the case reports presented by Deutman and Grizzard, the presence of fibrotic macular scars and subretinal haemorrhage are highly suspicious of previously existing subretinal neovascularisation. It is important not to

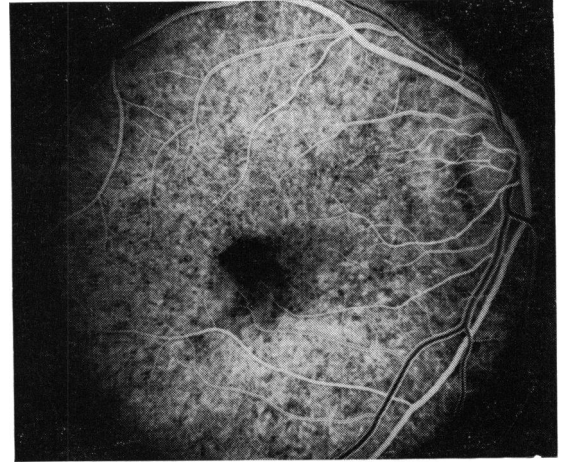


Fig. 3d *The laminar phase of the angiogram shows increased blockage of fluorescence in the macula due to subsensory retina and subretinal pigment epithelial haemorrhage.*

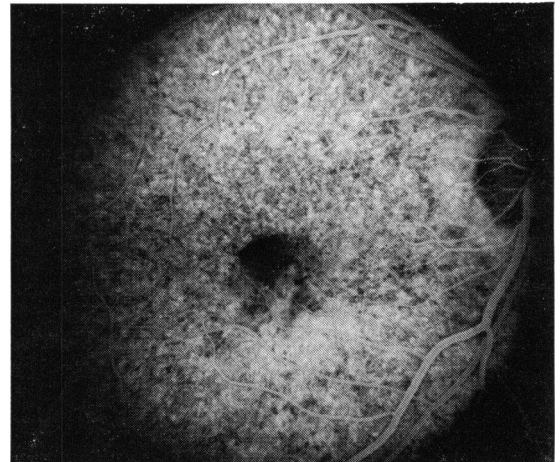


Fig. 3e *Venous phase of the angiogram continues to show blockage of the background fluorescence. There is an area of increased hyperfluorescence due to fluorescein leakage presumably from a subretinal neovascular membrane.*

attribute automatically a decrease of visual acuity in rubella patients to cataracts or amblyopia. It is necessary to look diligently for a macular lesion. Subretinal neovascularisation and macular haemorrhage result in fibrotic macular scars, which lead to a significant reduction in central acuity.

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