Correspondence

Vitelliform degeneration and butterfly-shaped pigment alterations

Sir, I read with interest Gutman et al.'s article on vitelliform macular dystrophy (VMD) in the right eye and butterfly-shaped epithelial dystrophy (BED) in the left eye of a 22-year-old female. They suggested that these were both expressions of one and the same disease. They referred to the concept of 'patterned' or 'pattern' dystrophies of the retinal pigment epithelium (RPE) by Hsieh et al. and Marmor and Byers, in which publications reticular pigment alterations, butterfly-shaped and granular pigmentations occurred in 2 families on a presumed autosomal dominantly inherited manner. These last 2 publications did not mention the simultaneous occurrence of VMD and BED in one family, or even any similarity, as is erroneously stated by the authors.

Both Best's VMD and Deutman's BED are autosomal dominantly inherited dystrophies of the RPE and both may show severely depressed EOG ratios. Adult onset vitelliform lesions without a diminished EOG ratio and without any detectable heredity have been described only recently. They are considered separate entities, different from Best's vitelliform degeneration. Deutman's butterfly-shaped foveal dystrophy is certainly another well-circumscribed nosological entity, which can be mimicked by several totally unrelated diseases: Stargardt's macular degeneration, acquired drusen of Bruch's membrane, rheumatic foveal degeneration, and myotonic dystrophy.

We had already demonstrated in a patient with an adult-onset vitelliform macular lesion in the same eye a butterfly-shaped degeneration of the fovea, as Deutman mentioned before us.

I consider any pattern or patterned degeneration of the RPE as a totally aspecific response of the RPE to very different pathogenic stimuli. We could also demonstrate reticular pigment alterations of the posterior pole of the eye in a patient with choroidal folds, and in another with multiple RPE detachments. A pattern(ed) dystrophy (which by its name implicates a hereditary disease) of the RPE does not exist, but there is certainly a pattern(ed) syndrome of the RPE, initiated by several different stimuli. So it is possible to find butterfly-shaped degenerative lesions of the RPE in patients with vitelliform disease of the RPE, even in these vitelliform lesions themselves.

There is no necessity to postulate a continuum or a common denominator for Best's VMD and Deutman's BED. Butterfly-shaped alterations can occur in a number of totally different RPE diseases, and possibly the same holds true for vitelliform lesions. There is no need either for the acceptance of the concept of pattern(ed) dystrophies of the RPE as a separate nosological entity. The existence of such a disease is the more unlikely because of the lumping together of autosomal recessive diseases (reticular and macoreticular RPE dystrophy), as well as autosomal dominant diseases of the RPE (Deutman's BED and fundus pulverulentus). However, the presumed occurrence of autosomal dominant reticular and macoreticular degenerations of the RPE adds to the confusion in designing a classification of the RPE diseases.

Nieuwe Bosscheweg 17, 5017 JJ Tilburg
Netherlands

References


Obituary

Jean M. Dollar, MS, FRCS, DOMS

Jean Dollar died on 20 April 1982 peacefully at home. She was born in London at the turn of the century. Her father, uncle, and grandfather were veterinary surgeons practising in Bond Street. After schooling in London she attended the London School of Medicine for Women and in 1926 graduated. She obtained the diploma in ophthalmic medicine and surgery in 1929, proceeding to her mastership in 1935 and final diploma of the Royal College of Surgeons in 1936. This same year she was appointed to the consultant staff of the Royal Eye Hospital, London. Her other consultant appointments included St Olave's, Elizabeth Garrett Anderson, and Royal Free Hospitals. In 1945 she was elected Hunterian professor at the Royal College of Surgeons of England.