LETTERS TO THE EDITOR

Differential diagnosis of iridocorneal endothelial syndrome and posterior polymorphous endothelial dystrophy

Editor,—In the paper by Laganowski et al.1 the point is made strongly that endothelial specular photomicroscopy (ESP) by revealing ICE cells and distinguishing between ICEb-PPD and ICEal-PPD, will always provide a definitive diagnosis even in uncertain cases and thus promote accurate management of patients and their families.1 Their own cases reflect the consistent unilaterality of iridocorneal endothelial (ICE) syndrome and the posterior bilaterality of posterior polymorphous dystrophy (PPD). The authors describe unilaterality in four out of 19 vesicular types, none out of two of the diffuse type and 14 out of 23 of the band type. Taken at face value this might suggest that there could be a large percentage of people with unilateral PPD disease that could be mistaken for the consistently unilateral ICE syndrome. On the other hand, the band type of PPD is so characteristic in its pattern that it does not share a similar slit-lamp picture with any ICE syndrome patient. Once these patients have been taken from the unilateral cases, then there is only one strictly unilateral case out of the 44 patients in whom a possible confusion could exist. Examination of this patient’s relatives would almost certainly show typical PPD findings which would confirm the diagnosis without the need for expensive instrumentation such as specular microscopy.

In addition, Laganowski et al.1 assert that subtotal ICE plus syndrome has a characteristic circumscribed nest of abnormal endothelial cells which is pathognomonic of ICE and not seen in PPD. Figure 11 in the paper by Hirst and Waring,2 bears a remarkable resemblance to Figure 2 of Laganowski et al.1 The patient described by Hirst and Waring2 came from a family of patients with posterior polymorphous endothelial dystrophy in which history confirmed the diagnosis in one of the family members. As Laganowski et al.1 contend ‘while the two conditions may conceivably co-exist in the same eye or in different members of the same family, the ICE syndrome and PPD are distinct entities,2 the only explanation for our case would be that the two diseases are co-existing in the same family, or perhaps it is more likely that the findings of Laganowski et al.1 are not pathognomonic of the subtotal ICE plus syndrome. Similar findings to these seen in Figure 2 were seen in a further 11 patients with the geographic and type of posterior polymorphous endothelial dystrophy in the original study by Hirst and Waring.1 Since that time, examination of further patients has confirmed these abnormal cells occurring in typical, familial and posterior polymorphous endothelial dystrophy.

In the first line of the discussion, it is suggested that Hirst and Waring reported the simultaneous occurrence of ICE syndrome and PPD in an individual or separately in different members of a family.2 We have not reported this. Perhaps the confusion was in the similar cell findings in some of our patients with geographic PPD and their patients with subtotal ICE plus. This finding is not ours alone as the similarity of cells as seen in Figure 2 of Laganowski et al.1 is also illustrated in Bourne’s1 wide field contact specular microscopy, a non-invasive, painless outpatient technique.

We assert that ‘ICE cells’, which give rise to the hammered silver appearance on slit-lamp examination and appear as a negative reverse endothelial line on specular microscopy, are pathognomonic of the ICE syndrome. In some cases of ‘subtotal ICE plus’ these cells may appear as an isolated feature in the remaining endothelium. We agree that Figure 11 in the paper by Hirst and Waring,2 resembles our Figure 2 of ‘subtotal ICE plus’. We contend that their Figure 11 shows classic ICE cells well demarcated from endothelium and is, therefore, different from the abnormalities quoted. This patient’s relatives have posterior polymorphous dystrophy (PPD) which would almost certainly confirm the diagnosis since we believe that the diagnosis of ICE or PPD can only be based on the clinical signs of the disease (providing, as this study has demonstrated, the condition has distinguishing features) and that ICE and PPD could co-exist in different members of the same family in the same eye.

Dr Hirst’s historical review of the ICE/PDD paper is from a relative of the case shown in his Figure 11. To extrapolate histological data from one family member with PPD to explain different physical signs in another member is scientifically unacceptable. Furthermore, this corneal button was not examined preoperatively by specular microscopy. The only currently practical means of deriving unequivocal evidence to distinguish ICE and PPD is to examine the endothelium from an affected eye with precise cell to cell correlation of specular photomicrographs with light and electron micrographs.

With regard to PPD, we disagree that examination of the corneal endothelium will not confirm the diagnosis. Moreover, in Hirst and Waring’s paper,3 the majority of relatives of patients with PPD showed no abnormality.

We disagree with Hirst’s statement that ICE is ‘consistently unilateral’; six of our 80 patients with ICE syndrome have typical ICE cells bilaterally. We do agree, however, given the advanced pathology of corneal buttons removed after penetrating keratoplasty that the histopathology may well represent a common pathway of cellular response and therefore be a poor discriminator of the primary diagnosis. We are sure that Dr Hirst would agree that the present enigma of ICE/PDD will only be resolved by close collaboration between clinician and pathologist, and preferably by building detailed comparative databases as a multicentre activity.

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