Letter to the Editor

DIDMOAD syndrome in a Chinese male with HLA DR7 DRw12

Sir,

Over 100 cases of the DIDMOAD syndrome (diabetes insipidus, diabetes mellitus, optic atrophy, deafness) have now been reported among Caucasians but it has not been previously described in the Chinese.¹

A 21 year old Chinese male with insulin-dependent diabetes mellitus for 11 years, and bilateral severe optic atrophy for 8 years presented with acute urinary retention. This was due to an atonic bladder and mega-ureters aggravated by diabetes insipidus. Additional features were high tone deafness detected by audiometry and a small pituitary fossa; anterior pituitary function was normal apart from a mild degree of primary hypogonadism. C-peptide secretion was severely impaired with a low fasting value of 0.04 ng/ml with no response to intravenous glucagon. There was no parental consanguinity and other family members were clinically unaffected and did not have diabetes insipidus or mellitus. His HLA type was, A3, A11, B15, B16, DR7 and DRw12.

Recent reports indicate an association of DIDMOAD with HLA DR2^{2,3} and though this antigen is present in 25% of the local Chinese population (Hawkins, B.R., personal communication), it was not present in our patient. Our finding is in agreement with the 2 cases described by Bertram *et al.*⁴

In DIDMOAD, the absence of islet cell antibodies,² persistence of C-peptide secretion⁵ (not seen in our patient), and absence of the DR3 antigen commonly associated with insulin-dependent diabetes mellitus (IDDM) supports the idea that diabetes in DIDMOAD may have a different pathogenesis. A recent review of 42 cases⁶ of optic atrophy and diabetes mellitus highlighted the absence of diabetic retinopathy which could perhaps be related to the different genetic basis of diabetes in DIDMOAD and in IDDM. However, it seems equally plausible that the lack of diabetic retinopathy is related to the altered metabolic climate in the retina secondary to optic atrophy.

Though the exact aetiology of DIDMOAD is not known, evidence to date points to an autosomal recessive progressive degenerative basis. It is an important syndrome to recognize,

as meticulous management of the urinary tract anomalies may prevent renal damage.

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