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Refractory Coeliac Disease in Diabetes Mellitus

Paul J Ciclitira

University of East Anglia, United Kingdom

Coeliac disease (CD, gluten-sensitive enteropathy) affects 1% of people in the EU and USA including 6% of subjects with type one diabetes mellitus. The pathogenesis of CD involves aberrant immune response, both adaptive and innate to gluten proteins from wheat and related cereals in the small intestinal mucosa of affected subjects. Refractory coeliac disease (RCD) is a complication of coeliac disease (CD) and involves malabsorption and villous atrophy despite adherence to a strict gluten-free diet (GFD) for at least 12 months in the absence of another cause. RCD is classified based on the phenotype of the T-cell morphology within the small intestinal intra-epithelial lymphocytes (IEL), into type 1 with normal polyclonal T cell receptors (TCR) of the IEL and type 2 with aberrant monoclonal TCR by PCR (polymerase chain reaction) for TCR at the β/γ loci. RCD type 1 is made do with strict nutritional and pharmacological administration.

RCD type 2 can proceed to ulcerative jejunitis or enteropathy associated lymphoma (EATL), the latter resulting in a 50% five-year mortality of subjects with RCD2. Management options for RCD type 2 and response to treatment vary between centres; there have been debates over the best treatment options. Therapy that have been utilized consists mycophenylate, azathioprine and steroids, cyclosporine, methotrexate campath and cladribine or fluadribine with or without autologous stem cell transplantation. We have treated RCD2 with prednisilone and azathioprine, replacing the latter with mycophenylate when there is azathioprine sensitivity in the treatment of RCD2. Our results employing prednisilone and azathioprine reveal a good response with histological recovery in 56.6% of treated individuals and without any mortality.

e: pciclitira@btinternet.com



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