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Early initiation of eculizumab in dense deposit disease case series and literature review

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Background: Dense deposit disease (DDD), a subtype of C3 glomerulopathy, is a rare disease secondary to hyperactivity of the alternative complement pathway with characteristic electron-dense deposits in the glomerular basement membrane on renal biopsy. Eculizumab, a monoclonal antibody prevent formation of the membrane attack complex through binding to C5 may be beneficial in its treatment. Reported cases are scarce, particularly in children.

Method: We report three pediatric patients with diagnosis of DDD on native kidney biopsy. All had acute kidney injury, hypertension, proteinuria >1 gram/day, and depressed level of C3. All Patients received methylprednisolone pulse therapy and Mycophenolate Mofetil. One patient required dialysis. Eculizumab was added early in two patients and in the third patient initiated after being on dialysis.

Results: In two patients, in whom Eculizumab started early, proteinuria and renal function improved significantly within weeks of treatment.C3 has been normalized in one patient and still depressed in the other. In the third patient who required dialysis, there was no response, she went into end stage renal disease (ESRD) and maintained on dialysis.

Conclusions: Eculizumab treatment was associated with reduction in proteinuria and improvement in renal function in two patients. Our findings are in agreement with previous reports of Eculizumab beneficial effect in DDD. Additionally, early initiation of Eculizumab may prevent progression to ESRD.

Biography

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