

that blocks the cleavage of C5 and impedes the generation of potent anaphylatoxin C5a and the formation of C5b-C9. It can be considered as a good treatment option for patients with atypical HUS associated with defects in both soluble and membrane bound complement regulators. The existing data on patients with atypical HUS in children is quite promising. Several reports of the successful use of monoclonal antibodies in patients of atypical HUS regardless of identified mutations or other risk factors, suggests that chronic Eculizumab / rituximab therapy might provide meaningful clinical benefit as measured by the resolution of thrombotic microangiopathy. Our own experience has also been encouraging.

Supportive therapy includes maintenance of fluid and electrolyte balance, adequate blood-pressure control, prophylactic antiepileptic in patients with neurologic symptoms, control of azotemia, optimize nutrition and monitor renal function and hemodynamics.

Complications include renal failure, stroke, coma, seizures and bleeding complications

Prognosis

- Patients collectively have a poor prognosis, and as many as 50-60% progress to ESRD (50% in those with the sporadic forms and 60% in those with the familial forms) or develop irreversible brain damage. About 25% die during the acute phase.
- The recurrence rate in patients receiving renal transplants is as high as 50%, with graft loss occurring in more than 90% who have recurrence. Recurrence rates are higher in patients with HF1 mutation.

CONCLUSION

Atypical haemolytic uremic syndrome (aHUS) is a thrombotic microangiopathy; occurs rarely; and comprises of triad of microangiopathic haemolytic anaemia, thrombocytopenia, and acute kidney injury. Recent studies show abnormalities in the mechanisms complement regulation as causes of aHUS. The prognosis is very poor, with the first aHUS attack being associated with a mortality rate of approximately 25%, and with approximately 50% of cases resulting in end-stage renal disease requiring renal replacement therapy.

This case highlights an uncommon presentation of a rare disease which needs a very high index of suspicion keeping in mind the very high morbidity and mortality associated with it.

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