

RARE PRESENTATION OF ATYPICAL HEMOLYTIC UREMIC SYNDROME(aHUS) AS RENOMEGALY TREATED SUCCESSFULLY WITH ECULIZUMAB

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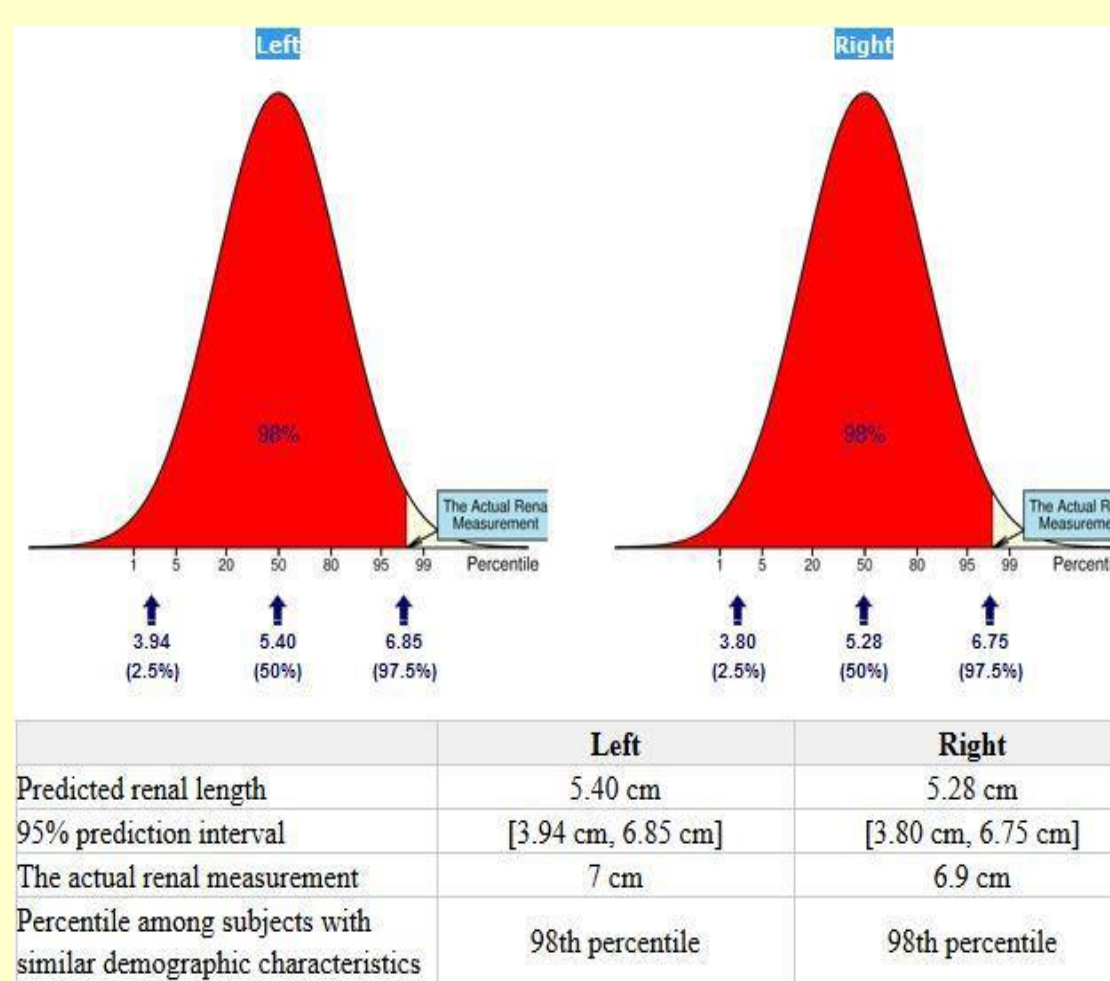
Introduction:

Atypical Hemolytic Uremic Syndrome(aHUS) is a disorder of complement system which can have varied presentation. Renomegaly is one of the manifestation of aHUS which is often underemphasized.

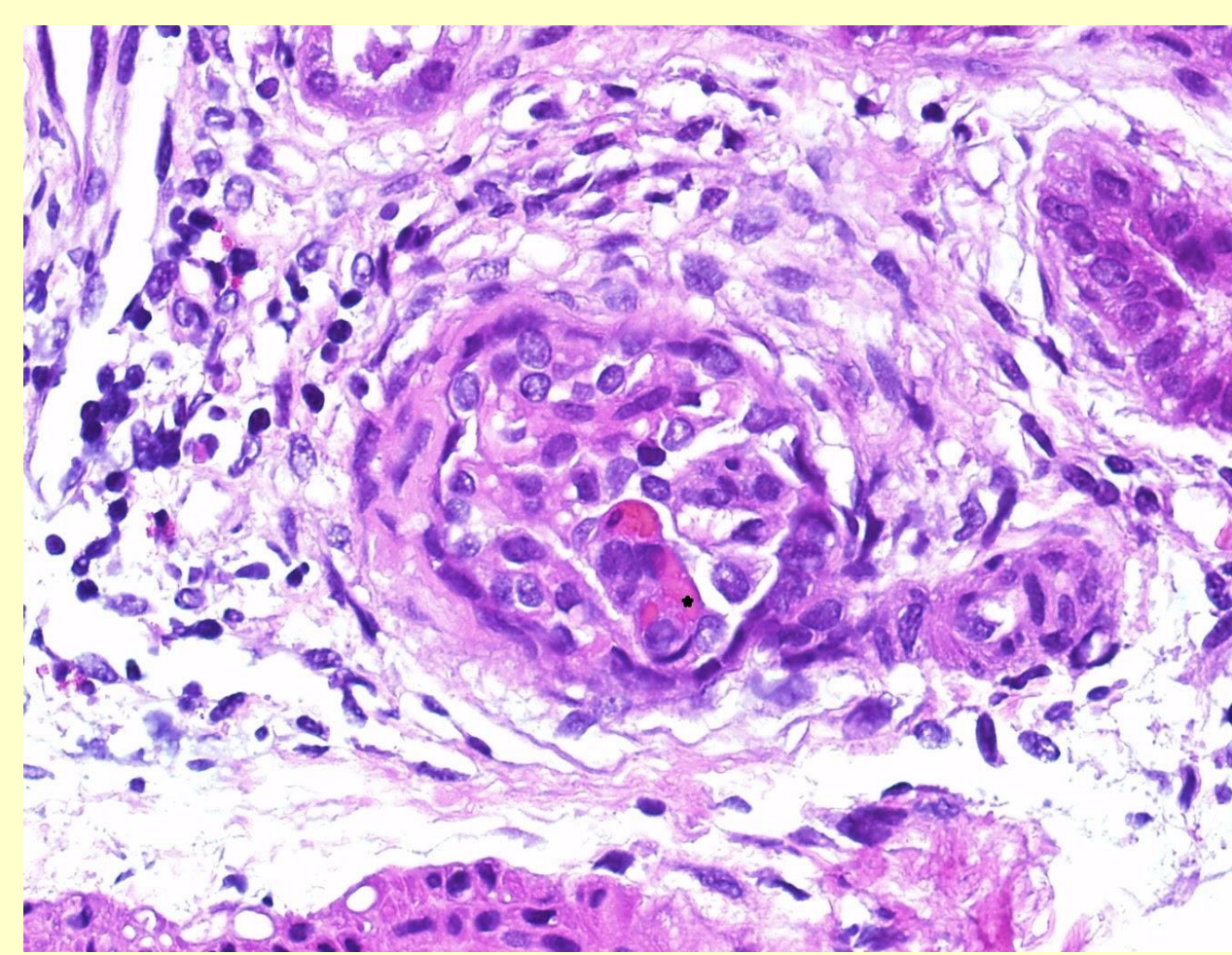
Case Report:

We report the case of a 7 month old male child who presented at 7 wk of life with respiratory distress and had features of anemia, acute kidney injury and hypertension. Platelet count was normal and LDH was increased. Renal imaging showed bilateral renal sizes 6.8 and 6.7 cm with increased echogenicity. He required multiple blood transfusions and antihypertensives. A renal biopsy was done which showed features of thrombotic microangiopathy(intracapillary thrombus in slide 1 and mesangiolytic changes(intracapillary thrombus in slide 1 and mesangiolytic changes/capillary wall splitting in slide 2) He was initiated on Eculizumab(Ecu) which improved his renal and hematological parameters. Genetic testing did not reveal any mutation.

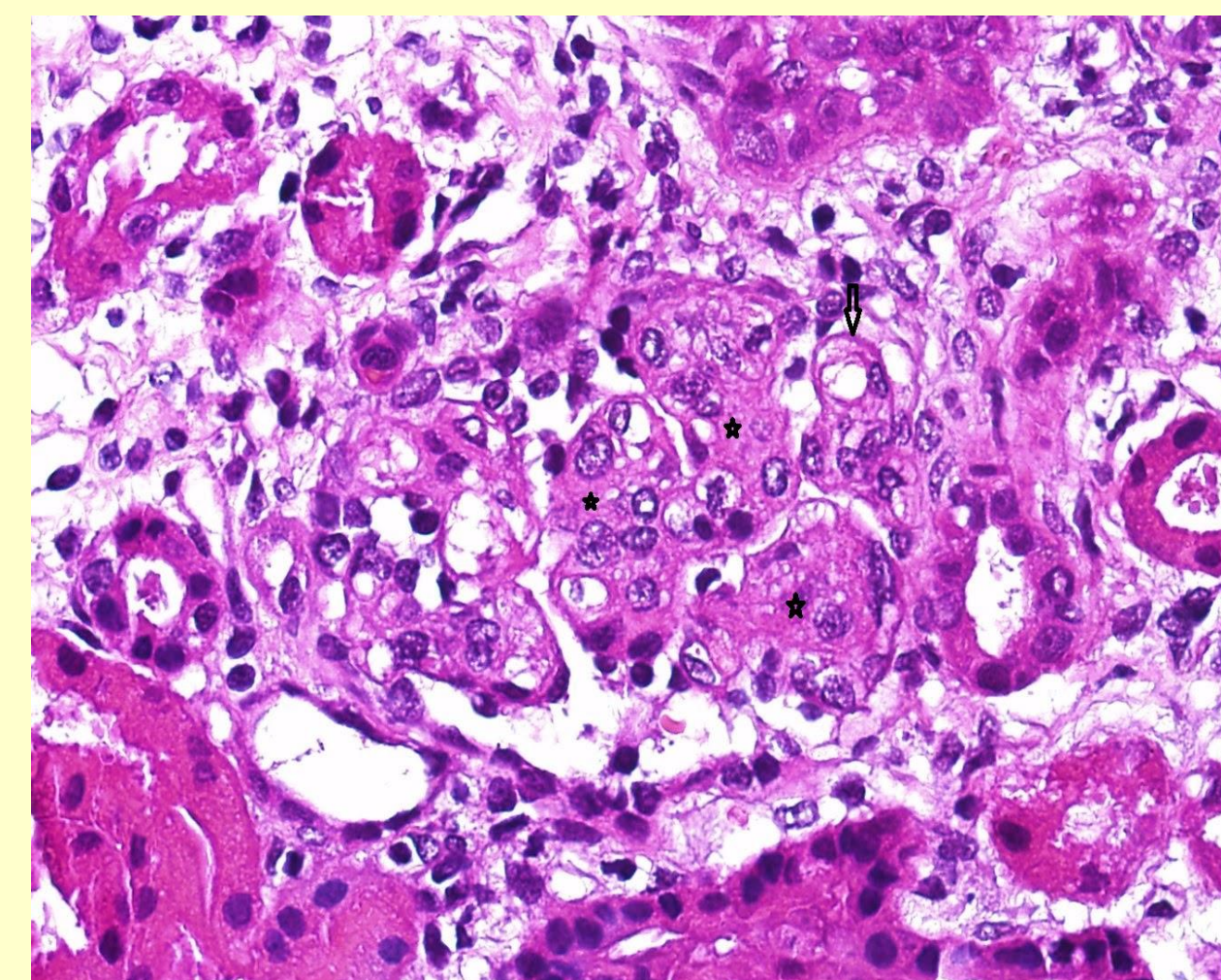
Graphs and tables



Renal size centiles at 7 week of age



Renal biopsy showing intra capillary thrombus



Renal biopsy showing mesangiolytic changes ("moth-eaten appearance", stars) and segmental splitting of capillary walls ("double contour", arrow)

LABORATORY PARAMETERS		
PARAMETER	PRE- ECULIZUMAB	POST ECULIZUMAB
HEMOGLOBIN	59 g/L	105 g/L
PLATELET	259,000/cu mm	282,000/cu mm
CREATININE	91 micromol/L	25 micromol/L
UREA	11 mmol/L	4 mmol/L
LDH	956 IU/l	356 IU/l
HAPTOGLOBIN	<0.10 g/L	<0.10 g/L
RETICULOCYTE COUNT	5%	0.5%

Results:

Child showed improvement in clinical and laboratory parameters on Ecu.

Conclusions:

aHUS should be considered in the differential diagnosis of renomegaly especially if any marker of thrombotic microangiopathy is positive in a hypertensive child.

References:

- Chen JJ, Zhi J, Mao W, Steinhardt GF. MrNomogram: a web-based multivariable pediatric renal nomogram. J Pediatr Urol. 2006 Oct;2(5):436-8.
- Kenney PJ, Brinsko RE, Patel DV, Spitzer RE, Farrar FM. Sonography of the kidneys in hemolytic uremic syndrome. Invest Radiol. 1986 Jul;21(7):547-50.

