May 4th, 11:30 AM - 1:30 PM

Diagnostic Features and Clinical Outcomes of Children with Tubulointerstitial Nephritis and Uveitis Syndrome (TINU)

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Lay Summary

TINU is a rare condition that causes inflammation of the kidneys (**T**ubulo**I**nterstitial **N**ephritis) and eyes (**U**veitis). This research study describes 9 children with TINU. We found that a urine test called beta-2-microglobulin (β2MG) can detect kidney inflammation in some children that might have otherwise not been recognized with usual clinical testing. All patients had eye inflammation that was hard to control. With treatment, all had good vision but 4 developed mildly decreased kidney function.

Background/Objective

Tubulointerstitial nephritis and uveitis (TINU) syndrome is an inflammatory disease that affects the kidneys and eyes. TINU is rare, but likely under-recognized due to variable presentation, nonspecific symptoms, and lack of universally accepted diagnostic criteria. The optimal treatment approach is unknown. This study aimed to contribute further knowledge about TINU by describing disease features and treatment response in a single-center cohort.

Methods

A retrospective chart review was performed of children diagnosed with TINU at a tertiary care children's hospital since January 2013. Demographics, clinical features, laboratory testing, eye exam findings and complications, kidney biopsy results, and treatment response are described.

Results

Table 1. Subject Demographic and Clinical Characteristics

Characteristic	N (%) or mean (range)
Age at diagnosis (years)	12.4 (8.8-16)
Male	6 (66.7%)
Presenting disease feature	
Uveitis (TIN diagnosed during initial workup)	6 (66.7%)
Uveitis (TIN diagnosed during medication taper at month 51)	1 (11.1%)
Tubulointerstitial nephritis (uveitis diagnosed during initial workup)	2 (22.2%)
Baseline Laboratory Findings	
Elevated ESR	3 (33.3%)
Anemia	6 (66.7%)
Elevated serum creatinine	5 (55.5%)
Baseline urinalysis findings	
Normal	3 (33.3%)
Trace glucose or trace protein only	2 (22.2 %)
More significant abnormalities (proteinuria, hematuria, and/or glycosuria)	4 (44.4%)
Urine β2MG	
Elevated at baseline (normal < 300 mcg/L)*	8 (88.9%)
Mean β2MG	14,434 (764-47,500 mcg/L)
Impaired final estimated glomerular filtration rate (eGFR) <90mL/min/1.73m2)	4 (44.4%)
Uveitis characteristics	
Bilateral	7 (77.8%)
Symptomatic	8 (88.9%)
Isolated anterior uveitis	5 (55.5%)
Anterior uveitis with papillitis (optic nerve edema)	4 (44.4%)
Ocular complications	
Synechiae	5 (55.5%)
Cataract	2 (22.2%)
Band keratopathy	1 (11.1%)
Best corrected visual acuity (BCVA) better than 20/40	n=18 eyes
Baseline	14 (77.8%)
Final	18 (100%)

*One patient did not have β2MG checked at presentation- β2MG was later elevated at time of uveitis flare/AIN diagnosis.

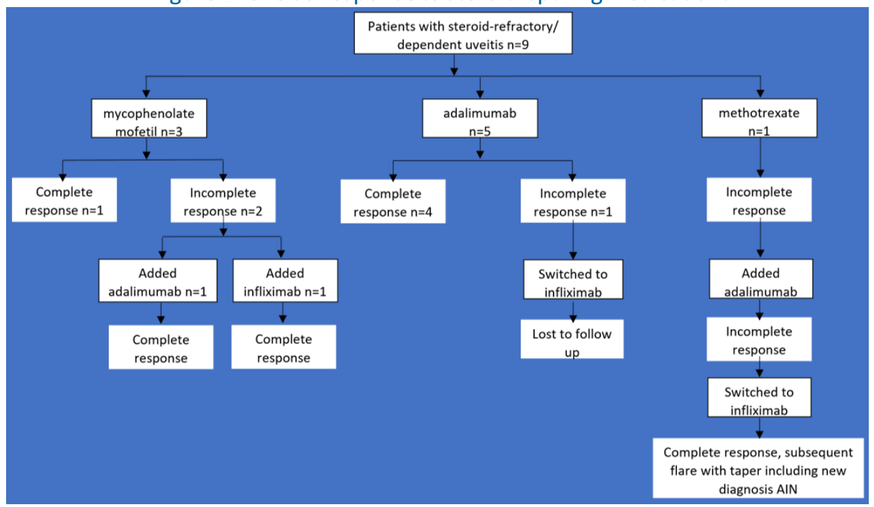
Clinical features are shown in Table 1.

- 100% patients demonstrated uveitis at initial diagnosis.
- 4 patients had normal serum creatinine and 5 had normal or near normal urinalysis; however, urine β2MG was significantly elevated in all patients at the time of AIN diagnosis.

Treatment response is shown in Figure 1.

- 100% required at least one chronic immunomodulator for uveitis control.
- 8 patients achieved uveitis quiescence, one was lost to follow up. 5 had at least one ocular complication, but visual outcomes were good (best corrected visual acuity 20/40 or better in 100%) at last visit.
- All had improvement in eGFR, with 5/9 (55.5%) normal at the last visit.

Figure 1. Uveitis response to steroid-sparing medications



Conclusions

- Urine dipstick and serum creatinine alone were inadequate to identify all patients with AIN; however, urine β2MG may be a more sensitive screening tool for TINU in children with uveitis.
- All patients required chronic immunomodulation to achieve steroid-free uveitis control; most required tumor-necrosis-factor-α inhibition.
- Future multi-center studies are key to furthering understanding of this rare condition and determining optimal treatment approach.

Disclosure: All authors have no financial interests or relationships to disclose.