Case report: A 28-year-old woman presented at the Emergency Department because of fever up to 39.5°C since 11 days together with flu-like symptoms. Blood tests revealed an hepatitis and 14% of atypic lymphocytes. Both the HIV and CMV serology were suggestive of an acute infection based on the presence of p24 antigen and an undetermined immunoblot, on the one hand, and the presence of CMV IgM antibodies in the absence of IgG antibodies on the other hand. On day 13 of fever, she experienced a sudden and sustained severe pain in the right shoulder without signs of neurologic involvement at clinical examination. Anti-retroviral treatment (ART) comprising dolutegravir (50mg QD) and emtricitabine/TAF (200/25mg) was initiated on the same day. At day 6 of ART, patient reported to be apyretic since 4 days but together with the persistent right shoulder pain, hypo- and dysesthesia in the distal right arm were mentioned. At physical examination, a scapula alata (winging scapula) was observed together with an atrophy of the right shoulder muscles, an areflexia of the right arm as well as a dysesthesia and impaired discrimination at the radial side of the distal right arm, suggestive of a NA for which a conservative approach was taken. Baseline hiv-1 viral load was determined to be > 7 log_{10} copies/ml with $CD4^+$ T-cells at 1962/mm^3 (29%). Liver function tests normalized rapidly and patient seroconverted for CMV IgG antibodies. At 1 month after onset of symptoms, shoulder pain had resolved. The performed EMG was compatible with a fasciculus lateralis lesion with signs of m. biceps denervation, confirming the diagnosis of NA. Muscular paresis has not yet improved, 4 months after onset.

Conclusion: To the best of our knowledge, this is the first case report of NA in the context of both an acute HIV and CMV infection. Even though the approach is conservative, this case report indicates the interests of a correct and early diagnosis to avoid useless examinations and treatments.

0123 The many faces of atypical Hemolytic Uremic Syndrome (aHUS) in Belgium

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ABSTRACT

Objective: The Belgian section of the aHUS registry (NCT01522183, sponsored by Alexion, Inc.) included 78 patients as of APR/1/2018. It constitutes the largest Belgian database on aHUS. We, herein, report on local disease features.

Methods: *Eligibility*: any consenting patient with a clinical diagnosis of aHUS, regardless of age, time of disease onset or treatment. *Exclusion*: ADAMTS13 activity below 5% (if performed) and the detection of a Shiga toxin (if the only cause of HUS). Patients were recruited between 09/2013 and 03/2017 through Belgian academic centers. Importantly, patients concomitantly enrolled in concurrent phase 2 clinical studies could not participate. 'Pediatric' patients are defined as younger than 18 y at enrolment. Results are expressed as percentage or median and range.

Results: *Epidemiology*. The registry counts 56% of women ($n = 44$) and 82% of adults ($n = 64$). Among them, 22% ($n = 14$) have a positive familial history. An underlying condition was mentioned in 19% ($n = 15$: malignant hypertension (4), infections (6), systemic lupus erythematosus (1), disseminated intravascular coagulation (1), malignancy (1) or others (2)). Median ages at disease onset and diagnosis are 3.1 [0.1, 15] and 3.2 [0.1, 15] y in children and 36 [1.2, 76] and 36 [1.2, 78] in adults. Overall, the median age at disease onset is 28 y [0.1, 76]. The median follow-up time is 2.3 y [0, 4.5]. *Disease burden*. As of APR/1/2018, end-stage renal disease concerns 34% of adults (n = 9/64 being dialyzed and 13/64 transplanted) and 29% of children (n = 3/14 being dialyzed and 1/14 transplanted).

Treatment by eculizumab: As of APR/1/2018, 44% of adults (n = 28/64) and 57% of children (n = 8/14) have already been treated by eculizumab. Eighteen adult patients and 5 children are still treated. Concerning safety: one meningoencephalitis caused by Neisseria meningitidis (serogroup Y) was reported under eculizumab. This infection occurred in a transplanted child vaccinated 5 years earlier and taking penicillin as antibiotic prophylaxis. He eventually recovered without sequela.

Causes of aHUS

	Genetic variants				
	CFH	*CFI*	*CD46*	*CFB*	*C3*
All, *n* #	15 (13 + 2)	2 (2 + 0)	11 (8 + 3)	1 (1 + 0)	4 (4 + 0)
All, *% among those tested*	24	3.6	19	2.5	9
N tested for the condition	62	55	58	40	44
Dialyzed, *n*	4	0	3	0	0
Transplanted, *n*	4	0	0	0	0

				Anti-CFH antibodies	Underlying Systemic disease
THBD	*DGKe*	*combination*[a]	*Not found*[b]		
1 (0 + 1)	3 (1 + 2)	4 (3 + 1)	22 (17 + 5)	7 (6 + 1)	15 (11 + 4)
3.2	11	5	28	12	19
31	28	78	78	60	78
0	0	2	3	1	1
0	0	0	0	1	1

[a] Patients with at least 2 pathogenic mutations
[b] If at least 5 genes tested but none of them was pathogenic
(n + n) denotes adult + pediatric patients

Conclusion: Belgium aHUS registry data support the fact that aHUS is a severe disease concerning both adult and pediatric populations. The wide variety of etiologies found advocate for a large panel of causes to be investigated in each patient with aHUS. Disease genetic background appears very similar to that of France[1] but notably, one Thrombomodulin-related case of aHUS is reported. Also, CD46 variants are more often recorded. Complement factor H variants were the most prevalent genetic causes among aHUS patients requiring renal replacement therapy. Finally, aHUS diagnosis and treatment require highly demanding management and resources and should be performed in specialized centers.

Reference

[1] Loirat et al. Pediatr Nephrol. 2016;31:15–39

0011 Investigating the Role of the Gut Microbiome in the Inflammatory State of Myeloproliferative Neoplasms

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