irAEs	n	frequency	Time (months)	Treatment	Discontinuation of immunotherapy	Response to treatment
Gastrointestinal tract	17	13.6%	4	23% steroid	58% no	17% steroid
Colitis	8			5% Infliximab	29.4% definitive	83% improvement
Intestinal perforation	1			72% nothing	11.7% temporary	
Hepatitis	8					
Endocrine glands	11	8.8%	4	18% steroid	90% no	63% stable
Hypophysitis	1			82% Other	10% definitive	37% improvement
Hypothyroidism	7					
Adrenal insufficiency	2					
Other	1					
Skin	10	8%	4	20% steroid	60% no	100% improvement
Rash/Prurito	7			20% topic treatment	30% definitive	
Vitiligo	1			60% other	10% temporal	
Other	2					
Rheumatological	7	5.6%	5	20% steroid	72% no	100% improvement
Arthralgias	3			20% local treatment	28% definitive	
Polymyalgia rheumatic	1			60% other		
Sicca syndrome	1					
Inflammatory myopathy	1					
Tenosynovitis	1					
Haematological	4	3.2%	Not available	25% steroid	50% no	25% death
Thrombocytopenia	2				25% definitive	75% improvement
Neutropenia	1				25% temporal	
Pancytopenia	1					
Lung (Pneumonitis)	3	2.4%	12	100% steroid	66% definitive	33% stable: 33% improvement; 33% death
					33% temporary	
Ophthalmologic	1	0.8%	Not available	Not available	Not available	Not available
Neuropathy	1	0.8%	2	steroid	Not available	Improvement
Cardiac	1	0.8%	Not available	Not available	Not available	Not available

THU0632

IDENTIFICATION OF ABERRANT T-CELL PHENOTYPE IN EPISODIC ANGIOEDEMA WITH HYPEREOSINOPHILIA (GLEICH SYNDROME) IN 30 PATIENTS: FREQUENCY, CLINICAL IMPLICATION AND PROGNOSIS

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Background: Gleich's syndrome or "Episodic Angioedema with eosinophilia" (EAE) is a rare disorder characterised by recurrent episodes of angioedema, concomitant hypereosinophilia and frequent elevated serum Immunoglobulin M (IgM) levels. So far, no series were reported in literature.

Objectives: In this retrospective French study we report the clinical spectrum and therapeutic management of patients with EAE in a large French nationwide retrospective multicenter cohort study, with a particular focus on lymphoid variant of hypereosinophilic syndromes-related EAE.

Results: Thirty patients were included with a median age at diagnosis of 41 years⁵⁻⁸⁴ and a median follow up duration of 53 months.³¹⁻⁹⁹ The median time between two angioedema crises was 30 days⁷⁻¹⁹² and the median duration of each crisis was 5.5 days¹⁻⁹⁰ with swelling affected the face, the neck, the upper limbs, and the lower limbs. Twelve patients (40%) had an abdominal. The median blood eosinophil count at diagnosis was 6.9 G/L [1.44-53.5] and total serum IgM levels were increased in 20 patients (67%). Abnormal T-cell immunophenotypes were detected in 12 patients (40%): CD3-CD4+ in 8 patients, CD3 +CD4+CD7 in 2, CD3-CD3ic+CD4 CD8-CD5+CD7 in 1 patient and CD3 +4-8-2+5+7+TCR γδ in 1 patients with presence of clonal TCR γ gene rearrangement in 5 cases (17%). Fourteen patients (47%) had bone marrow cytology and/or histology revealing high eosinophil counts and/or infiltration by non-dystrophic eosinophils in all cases. Eosinophilic perivascular infiltration (without vasculitis) was reported in 2 patients (7%) in which a skin biopsy was performed. An eosinophilic infiltrate was also found in various organs (stomach, gallbladder, liver, ileon, and colon) that were biopsied and/or surgically removed.

All 30 patients were initially treated with glucocorticoid with a median initial dose of 40 mg of daily prednisone.⁵⁻⁸⁰ After first line treatment and a median time of 15.2 months (95% CI: 9.4-50.1), disease was considered as uncontrolled in 24 patients (80%) with relapses in 15 patients (50%) or glucocorticoid's dependence in 9 (30%).

Glucocorticoid-sparing agents were used in 14 cases (47%) for relapses (n=8) or glucocorticoid- dependence (n=6). Associated treatments consisted of interferon-α (n=8, 27%), mepolizumab (n=2, 7%), ciclosporin A (n=5, 17%), methotrexate (n=4, 13%), hydroxycarbamide (n=2, 7%).

The presence of an abnormal T-cell population was the sole factor associated with a shorter time to relapse (hazard ratio 4.15 [CI 95% 1.18-14.66; p=0.02].

Two deaths occurred during follow-up (7%) due to lymphoma evolution for one and unexplained sudden death for the other.

Conclusions: EAE could be classified as a lymphoid variant of hypereosinophilic syndrome but that it is an heterogeneous condition. Although patients usually respond well to glucocorticoids, those with a circulating T-cell clonal population are at higher risk of both relapse and lymphoma suggesting necessity to long-term treatment and close monitoring.

Disclosure of Interest: None declared

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THU0633

ACUTE FORM OF SARCOIDOSIS (LÖFGREN SYNDROME) IN RHEUMATOLOGY PRACTICE

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Background : Löfgren syndrome (LS) is the acute form of sarcoidosis, characterised by erythema nodosum (EN), joints involvement, fever and intrathoracic lymphadenopathy. LS clinical polymorphism leads to diagnostic errors.

Objectives: To study the clinical, laboratory and radiological features of early stages of sarcoidosis in a cohort of patients referred to rheumatology centre.

Methods: The study included 125 patients (104 females and 21 males, mean age 42±12 y) with clinical and radiological features of LS. All patients were referred to rheumatology centre with EN diagnosis. The median duration of disease was 1 (0.5-2.0) months. All patients were subjected to comprehensive clinical and laboratory-instrumental examination, including biochemical panel and immunological parameters, chest X-ray or CT, as well as (15 cases) histopathology examination of nodular biopsy specimens of the skin and subcutaneous fat.

Results: In 97% pts EN involved the lower legs, mostly the anterior surface, in 35% - the hips, in 25% - the upper limbs and in 3% - the trunk. Symmetric lesions were documented in 50% of patients. Confluence of nodules forming conglomerates was registered in 48% pts. The involvement of more than 50% of lower leg surface (68%) was directly associated with the number of nodules (p<0.001; r=0.60) and C-reactive protein level (p=0.006; r=0.38). There was a direct correlation between the number of nodules and EN duration (p=0.04; r=0.20), and also the tendency to nodules confluence (p<0.001; r=0.39). Joint involvement was documented in 106 (85%) patients, with predominantly ankle (66.4%) and knee (40%) joints affected. Asymmetry of joint involvement was documented in 8% pts. Articular manifestations preceded the emergence of EN in 35 (28%) patients by mean 12±9 days. There was a significant direct correlation between the duration of articular syndrome and appearance of nodules and their number (p=0.02; r=0.45). Joint involvement after EN manifestation – in an average 12±7 days – was documented in 10% of cases. Male gender (odds ratio (OR) 6.5; confidence