Methods: PM/DM patients meeting the Bohan and Peter diagnostic criteria from the First Affiliated Hospital of Zhengzhou University were admitted between 2015 August to 2017 December. A retrospective analysis of the WBMRI imaging data, serum creatine kinase, electromyography and muscle biopsy were performed. The WBMRI images were estimated by semiquantitative score and the disease activity was estimated by MDAAT. SPSS 20.0 was utilised to analyse the statistical significance.

Results: This study includes 61 patients, including 41 DM cases and 20 PM cases, all of these shows typical pathological feature and inflammatory muscle in WBMRI. Significant statistical correlation was found between the grade of muscle oedema estimated by WBMRI and the clinical assessment through MDAAT for muscle disease activity (DM $r=0.57$, $p=0.006$; PM $r=0.84$, $p=0.001$). The positive rate of serum creatine kinase test was 63% (39/61). Besides muscular changes, we also detected 35 cases in interstitial lung disease, 2 cases in osteonecrosis, 1 case in bone marrow oedema.

Conclusions: WBMRI is a sensitive, noninvasive method to evaluate the disease activity, which is better than serum creatine kinase. And it is of great value to screen the complication or concomitant diseases such as ILD and osteonecrosis.

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THU0630

IMMUNE-RELATED ADVERSE EVENTS ASSOCIATED WITH IMMUNE CHECKPOINT INHIBITORS IN SPANISH PATIENTS

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Background: Immune checkpoint inhibitors (ICI), a type of immunotherapy which block negative co-stimulation of T-cells, have surfaced as an important alternative of treatment for advanced malignancies. Unfortunately, these agents have been associated with immune-related adverse events (irAE).

Objectives: To analyse the frequency, type and outcome of irAE in Spanish patients treated with ICI.

Methods: We carried out a retrospective, observational and longitudinal study, including adult patients diagnosed with cancer and treated with ICI followed in our centre (Spanish university hospital) from October 2012 to December 2017. Demographics, clinical, therapeutic and outcome data were collected from their medical records.

Results: We included 125 patients, 66.4% males, all them Caucasians, with a mean age at diagnosis 63 years (range:42-81). The most common types of cancers were lung carcinoma (61.6%) and melanoma (12.8%). The ICI most frequently used was nivolumab (29.6%), followed by pembrolizumab (8.8%) and atezolizumab (8.8%), as monotherapy. It was also common, combinations, or the use of consecutive way with different drugs. Occurrence of irAE was reported in 51 patients (table 1) and the systems more frequently involved were gastrointestinal tract, endocrine glands and skin. Most of these cases were treated steroids at high doses but a case of colitis required infliximab. Withdrawal of immunotherapy was required in almost half of the adverse events (45%). During the follow-up period there were 5 deaths associated with irAE.

Conclusions: The frequency and type of irAE associated ICI in our series were similar to those reported in previous studies, except for rheumatological events, being arthritis and sicca syndrome the most frequently reported in other series. These adverse events are not rare and may be a potential cause of morbidity in these patients, so it is important to recognise them early and treat properly, with the rheumatologist being one of the specialists involved.

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THU0631

REFRACTORY AND SEVERE UVEITIC CYSTOID MACULAR OEDEMA IMPROVES WITH TOCILIZUMAB IN DIFFERENT IMMUNE-MEDIATED INFLAMMATORY DISEASES

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Background: Cystoid macular oedema (CME) represents the leading cause of blindness in uveitis of different immune-mediated inflammatory diseases (IMIDs).

Objectives: Our aim was to evaluate the efficacy of Tocilizumab (TCZ) in different IMIDs with refractory CME.

Methods: Multicentre study of 24 patients with CME due to uveitis of different IMIDs refractory to traditional treatment with systemic corticosteroids and at least one conventional immunosuppressive drug including in most cases biological therapy (n=21). CME was defined by (OCT >300 µm). We studied CME with TCZ in 4 different IMID: juvenile idiopathic arthritis (JiA), Behçet's disease (BD), Birdshot retinochoroidopathy (BR) and idiopathic.

The main outcome was the improvement of macular thickness. Other variables assessed were inflammation of the anterior chamber and vitreous and best corrected visual acuity (BCVA)

Results: We studied 16 ♀/8 ♂, mean age 35.2±19.3 years. The associated diseases were: JiA (n=9), BD, BR⁴ and idiopathic.⁴ The ocular patterns were: panuveitis,⁹ anterior uveitis,⁶ posterior uveitis⁵ and intermediate uveitis.⁴ Most patients had bilateral involvement.²² The biological therapy used before the administration of TCZ were infliximab,⁸ adalimumab,¹⁸ etanercept,² golimumab,² rituximab,² abatacept,³ anakinra¹ and dactilizumab.¹

TCZ administration schedule was 8 mg/kg/4 weeks iv.²³ or every 2 weeks.¹ TCZ was used in monotherapy¹² or combined with conventional immunosuppressive drugs.¹² OCT values improved considerably in 12 months: in JiA from 340.6±134.1 µm to 252.5±30 µm, in BD from 375.1±117 µm to 235±7.1 µm, in BR from 550.7±214.4 µm to 295.5±43.2 µm and in idiopathic from 515±219.6 µm to 208.3±46.7 µm (figure 1). Inflammation in anterior chamber and vitritis and BCVA also improved in the 4 subtypes. No minor side effects were observed, so no patient had to stop treatment.

Conclusions: TCZ seems a rapid effective treatment in severe and refractory uveitic CME, regardless of the underlying IMIDs.

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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 hyper eosinophilia 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 hyper eosinophilic 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 hyper eosinophilic 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.

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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