Table 1. Clinical characteristics of 24 pediatric SAPHO patients

Skin manifestation	**17/24 (71%)**
Severe acne (M/F)	11/0
PPP (M/F)	0/5
None (M/F)	3/4
Psoriasis (M/F)	0/1
Nonspecific rash (M/F)	1/0
Bone manifestation	**24/24(100%)**
Pain	24 (100%)
Swelling	17 (71%)
Restricted function	19 (79%)

Table 2. Effect of different treating choices for 24 pediatric SAPHO patients

Medications	**Remission**	**Partial response**	**No response**	**Total**
NSAIDs	3	3	0	6/24
Bisphosphonates	7	2	1	10/24
TNF-α antagonist	7	3	0	10/24
Glucocorticoids	0	1	0	1/24

Conclusion: This is the first Chinese cohort of pediatric SAPHO patients. Their bone lesions could be divided into 4 types, anterior chest wall, mandible, peripheral bones and spinal bones. We also provide evidence that bisphosphonate and TNF-α antagonist are useful for pediatric SAPHO treatment.

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Disclosure of Interests: None declared

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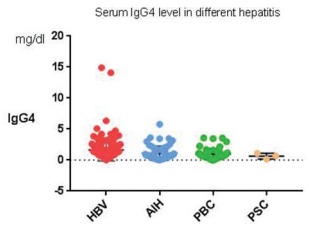
FRI0601 SERUM IMMUNOGLOBIN G4 LEVEL IN INHEPATITIS B WAS HIGHER THAN AUTOIMMUNE LIVER DISEASE IN A CHINESE POPULATION

Zetao Liao, Yanli Zhang. *Third Affiliated Hospital of Sun Yat-sen University, Guangzhou, China*

Background: High serum level of Immunoglobulin G4 (IgG4) was detected in autoimmune hepatitis (AIH), but little was known in other types of hepatitis such as hepatitis B, primary biliary cholangitis (PBC) and primary sclerosing cholangitis (PSC).
Objectives: The aim of this study is to evaluate the level of serum IgG4 in hepatitis B and autoimmune liver disease.
Methods: Hepatitis B and autoimmune liver disease patients were enrolled between January 1st and August 30th 2018. Diagnosis of Hepatitis B, AIH, PBC and PSC was basing on corresponding classification criteria, and established IgG4 related diseases were excluded. The levels of serum IgG4 were determined by nephelometric assay. The cut-off value of serum IgG4 was 2.01 g/L. Serum IgG4 among different liver diseases were compared.
Results: 161 HBV hepatitis and 112 autoimmune liver diseases (52 AIH, 57 PBC, 3 PSC) were enrolled in this study. The HBV hepatitis patients were younger than patients with autoimmune liver disease (44.79±12.56 vs 52.52 ± 11.16 years old). In total, 61 patients (22.34%) had elevated serum IgG4 level. IgG4 level in HBV hepatitis was significantly higher than in AIH, PBC, respectively (1.64±1.80 vs 1.11±1.16, 1.64±1.80 vs 0.96±0.87 g/L, P<0.05).



Conclusion: High serum IgG4 was very common in liver diseases without IgG4 related diseases especially in HBV hepatitis. Differential diagnosis should be made in patients with hepatitis accompanied with high serum IgG4.

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[1] No.

Disclosure of Interests: None declared

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FRI0602 SERUM ALBUMIN PREDICTS ONCOLOGICAL OUTCOMES BUT NOT RHEUMATIC IMMUNE-RELATED ADVERSE EVENTS FOLLOWING PD-1 INHIBITOR THERAPY

Christopher Mcmaster[1,2], David Liew[1,2,3], Bonnia Liu[1,2], Jessica Leung[1], Albert Frauman[2,3], Jonathan Cebon[4,5], Russell Buchanan[1,3]. *[1]Austin Health, Rheumatology, Melbourne, Australia; [2]Austin Health, Clinical Pharmacology and Therapeutics, Melbourne, Australia; [3]University of Melbourne, Medicine, Melbourne, Australia; [4]Olivia Newton-John Cancer Wellness and Research Centre, Melbourne, Australia; [5]La Trobe University, Cancer Medicine, Melbourne, Australia*

Background: Rheumatic immune-related adverse events (irAEs) following immune checkpoint inhibitor (ICI) therapy for cancer are an increasing burden on rheumatological services but have also been associated with subsequent good oncological outcomes(1). A clinical need for biomarkers to predict rheumatic irAEs exists. High serum albumin and low lactate dehydrogenase (LDH)(2) prior to ICI therapy has been associated with subsequent good oncological outcomes, and may therefore predict rheumatic irAEs, but to the best of our knowledge this has not previously been examined.
Objectives: To determine the relationship between serum albumin and LDH of immune-related adverse events (irAEs) and oncological outcomes following treatment with the main classes of ICI therapy for cancer, programmed cell death protein 1 (PD-1) inhibitors and programmed death-ligand 1 (PD-L1) inhibitors.
Methods: We retrospectively examined all patients at a single centre who had a serum albumin performed in the institutional laboratory before treatment with a PD-1 or PD-L1 inhibitor before January 1, 2018, with follow-up until October 1, 2018. Patients with any diagnosis of a rheumatic irAE were identified. Treatment response was determined based on assessment by the treating oncologist, with favourable oncological response defined as complete or partial response. Statistical tests were performed using two-tail t-tests. Missing laboratory values were imputed using median imputation.
Results: There were 401 episodes of therapy which met criteria, of which 324 had a serum albumin level and 244 had serum LDH prior to the commencement of therapy. A rheumatic irAE was diagnosed in 26 of the 401 patients (6.5%) and a good oncological response was noted in 140 patients (34.9%). There was a statistically significant relationship between higher baseline serum albumin and favourable oncological response ($p < 0.001$), but not with the development of a rheumatic irAE (p=0.41)(Table 1). Although serum LDH levels were lower in those with favourable oncological response, this relationship was not statistically significant (p=0.16). Similarly, LDH levels were lower on average in those who subsequently developed a rheumatic irAE, however this relationship was not statistically significant (p=0.57).
Conclusion: High serum albumin was associated with good oncological outcomes following ICI therapy with a PD-1 or PD-L1, but not with rheumatic irAEs. The reason for this dissonance is not clear. Not all markers of good oncological response predict rheumatic irAEs and further study should focus on consequent implications for rheumatic irAE pathophysiology.

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Table 1. Albumin and LDH, and associations to oncological response and rheumatic irAE development

	Oncological non-responders	Oncological responders	p-value
Albumin (g/L)	33.4	35.2	< 0.001
LDH (units/L)	262	237	0.16
	No rheumatic irAE	**Rheumatic irAE**	**p-value**
Albumin (g/L)	34.0	34.9	0.41
LDH (units/L)	254	238	0.57

Disclosure of Interests: None declared
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FRI0603 SARCOIDOSIS IN A TERTIARY HOSPITAL

Isabel Madroñal García, Clara Aguilera Cros, Maria Dolores Arcila Duran, Lara Mendez, Marina Gomez Vargas, Alberto Ruiz Roman, Ricardo Juan Gil Velez, Jose Antonio Rodriguez Portal, Noemi Patricia Garrido Puñal, Esteban Rubio Romero. *Hospital Universitario Virgen del Rocío, Reumatologia, Sevilla, Spain*

Background: Sarcoidosis (S) is a systemic granulomatous disease of unknown etiology. The most frequent affectation is pulmonary, ocular and cutaneous, although sarcoidosis can damage other organs, such as the musculoskeletal system.
Objectives: To describe the clinical characteristics and the radiological pattern, in a cohort with predominant pulmonary S, as well as to establish the relationship between the angiotensin converting enzyme (ACE) levels and the S course (chronification or remission).
Methods: This is a retrospective descriptive study of patients treated in our hospital, since 2008 to 2018, with diagnosis of S. The data was obtained through the review of medical records. The delay in the diagnosis of S was defined as the difference in months between the initial diagnostic suspicion and the final diagnosis of S.
We use Chi-square tests to study the association between ACE levels and the course of the disease.
Results: Fifty-five patients (31 women) were included, with a mean age of 52 ± 12 years. The first diagnosis was: 85% S, 10% lymphoma and 4% tuberculosis. The median of months for the definitive diagnosis of S was 5.5 months.
Extrapulmonary clinical manifestations in table 1.
The ACE is increased in 38 patients (70%). Simple x-ray and high resolution tomography of chest were done in all patients. Pulmonary stage 2 was the most frequent (51%), followed by stage 3 (16%), stage 0 (14%) and stage 4 (9%).
In 90% of the patients, histological confirmation was obtained by transbronchial (47%), cutaneous (11%) or lymph node biopsy (29%).
94% of the patients have been treated with oral glucocorticoids, 52% associate immunosuppressive therapy (Methotrexate 27% and Azathioprine 14%) or biological treatment (1 patient with Adalimumab and another with Infliximab). In 54% of the patients the S had a chronic course and in 43% S remitted.
Increased levels of ACE were associated with clinical remission of the disease and normal levels with chronicity (p: 0.013).

EXTRATHORACIC CLINICAL MANIFESTATIONS	N(%)
Skin	15(27)
Neurological	4(7)
Cardiac	8(14)
Renal	2(4)
Ocular	7(13)
Monoarthrittis	3(5)
Poliarthritis	6(11)
Bilateral swelling in ankles	6(11)
Hepatosplenomegaly	8(14)

Conclusion: The results of our study resembles, in general, what has been published in the literature. In our study, elevated ACE is associated with remission of the disease, contrary to the published, in which increased levels of ACE in symptomatic patients may reflect disease activity.
Disclosure of Interests: None declared
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FRI0604 SYSTEMIC SARCOIDOSIS. STUDY OF 381 PATIENTS FROM A TERTIARY UNIVERSITY HOSPITAL IN THE NORTH OF SPAIN

José Luis Martín-Varillas[1], Lara Sánchez Bilbao[1], Iñigo González-Mazón[1], Raúl Fernández Ramón[2], D. Prieto-Peña[1], David Martinez-Lopez[1], Eva Peña Sainz-Pardo[3], Belén Atienza-Mateo[1], Monica Calderón-Goercke[1], Rosalía Demetrio-Pablo[2], Vanesa Calvo-Río[1], Miguel A. González-Gay[1], Ricardo Blanco[1]. *[1]H.U.M. Valdecilla, Rheumatology, Santander, Spain*; *[2]H.U.M. Valdecilla, Ophthalmology, Santander, Spain*; *[3]H.U.M. Valdecilla, Paediatrics, Santander, Spain*

Background: Sarcoidosis is a systemic granulomatous disease characterized by the presence of non-necrotizing granulomas in different parenchyma.
Objectives: To describe demographic, clinical and analytical features in a cohort of patients with Sarcoidosis diagnosis from northern Spain during the last twenty years.
Methods: Descriptive study of 381 patients diagnosed with sarcoidosis during the period 01/01/1999 to 01/01/2019. Biopsy and/or clinical and compatible imaging tests were required for the diagnosis of sarcoidosis. Demographic parameters, clinical manifestations, complementary tests and treatment were registered. Results are expressed as mean±SD or as median and interquartile range (IQR) as appropriate.
Results: 381 patients (192 female/191 male, ratio 1:1), with a mean age 45.5±15.4 years at disease onset, and with 94.8% of Spanish nationality. 33% were smokers at diagnosis and 6.3% had tuberculosis (**Table 1**). The sarcoidosis incidence rate in our population was 3.3 cases/100000 p/year, similar to other national series[1].
Most frequent clinical manifestations were as follow: pulmonary symptoms (72.3%), general symptoms (37.3%), skin involvement (31%), joint manifestations (27.9%), ophthalmological manifestations (13.0%), digestive disorders (9.3%), neurological symptoms (6.5%), nephrological (4.7%) and cardiological involvement (1.6%) (**Table 1**). In addition, 11.5% of the patients had a Löfgren syndrome, 0.5% a Heerdfort syndrome and up to 11.5% had mediastinal adenopathy as an incidental finding in simple chest radiography.
A simple chest radiograph was performed in all patients. 83.7% presented abnormal patterns: stage I (41.4%), stage II (32%), stage III (7.6%) and stage IV (3.8%). In addition, 29.9% and 10.5% of patients presented pathological scintigraphy and PET respectively. Biopsy was performed in 81.9%, with a mediastinal approach in 43.3%. **(Table 2).**
After a median follow-up of 11.0 [6.0-17.0] years, we observed that up to 32% of patients never received treatment. In the remaining 68%, the main treatment was oral glucocorticoids with a mean dose of 43.4±19.1 mg/day. Other treatments used were conventional immunosuppressants and biological agents **(table 2).**

Table 1.

	Patients diagnosed with sarcoidosis (N=381)
DEMOGRAPHIC PARAMETERS	
Sex, n (%)	191♂/192♀ (50.1/49.9)
Age at disease onset (years), mean ± SD	45.5 ± 15.4
Current age (years), mean ± SD	59.0 ± 16.3
Follow-up, median [ICR]	11.0 [6.0-17.0]
Nationality (Spanish), n (%)	361 (94.8)
Smokers, n (%)	126 (33.1)
Asthma, n (%)	24 (6.3)
Tuberculosis, n (%)	24 (6.3)
Tumours, n (%)	23 (6.0)
CLINICAL MANIFESTATIONS	
Pulmonary symptoms, n (%)	**277 (72.3)**
- Dyspnea, n (%)	103 (26.9)
- Cough n (%)	35 (9.1)
- Pleuritic pain, n (%)	10 (2.6)
- Adenopathy, n (%)	129 (33.7)
Ocular involvement, n (%)	**50 (13.0)**
- Uveitis, n (%)	36 (9.4)
- VA, n (%)	5 (1.3)
- Exophthalmos, n (%)	2 (0.5)
- Others, n (%)	7 (1.8)
Skin involvement n (%)	**119 (31.1)**
- Erythema nodosum, n/N (%)	72 (18.8)
- Lupus pernio, n/N (%)	7 (1.8)
- Parotid gland hypertrophy, n/N (%)	7 (1.8)
- Granulomatous dermatitis, n/N (%)	33 (8.6)
Joint involvement, n (%)	**107 (27.9)**
- Arthralgia, n (%)	66 (17.2)
- Arthritis, n (%)	40(10.4)
- Sarcoidosis myopathy, n (%)	1 (0.3)
Digestive disorders, n (%)	**36 (9.3)**
- Hypertransaminasemia, n/N (%)	20 (5.2)
- Hepatic granulomas, n/N (%)	13 (3.3)
- Other granulomas, n/N (%)	3 (0.8)
Neurological symptoms, n (%)	**25 (6.5)**
- Headache, n (%)	13 (3.5)
- Meningitis, n (%)	2 (0.5)
- Neuropathy, n (%)	6 (1.5)
- Others, n (%)	4 (1.0)
Cardiological manifestations, n (%)	**6 (1.6)**
- Dilated cardiomyopathy, n (%)	4 (1.0)
- Pericarditis, n (%)	1 (0.3)
- SMVT, n (%)	1 (0.3)
Nephrological manifestations, n (%)	**18 (4.7)**
- Acute renal failure, n (%)	12 (3.1)
- Chronic kidney disease, n (%)	5 (1.3)
- Lithiasis, n (%)	1 (0.3)
Systemic symptoms, n (%)	**143 (37.3)**
- Fever, n (%)	36 (9.4)
- General syndrome, n (%)	107 (27.9)
Lymphadenopathy as incidental finding, n (%)	44 (11.5)
Löfgren syndrome, n (%)	44 (11.5)
Heerdfort syndrome, n (%)	2 (0.5)