The mean age at diagnosis was 45,01 years (SD 16,71 ± years), The median of tracing was seven yeas (P25 de four years/P75 de 14 years). The mean initial antiRNP titre was 239,33 the median was 144 (P25 58/P75 400). The initial diagnosis were Mixed Connective Tissue Disease 6,25% (4), Undifferentiated Connective Tissue Disease 31,25% (20), systemic erythematosus lupus 45,31% (29) and Others 14,06% (9). MCTD diagnosis was made in 29,69% (19) of patients at the end of follow-up. Patients with MCTD as final diagnosis presented in their first visit Raynaud phenomenon 73,68% (14), puffy hands 31,58% (5), acrosclerosis 10,56% (2), myositis 21,05% (4), Arthralgias 68,42% (13), arthritis 57,89% (11), Pulmonary arterial hypertension 5,26% and Interstitial lung disease 10,53% (2). The initial RNP titre was evaluated using the number of times over the laboratory cut point. The area under the ROC curve for the initial RNP was 0,70. Looking for the highest specificity with and adequate sensitivity, we selected the cut-point with highest LR +. The selected cut-point was ≥24,4 times the laboratory cut-point, with s specificity of 77,78 and a sensitivity of 57,89.

Conclusions: The cut-point for low/high RNP titres measured by ELISA in our cohort of patients with MCTD as a final diagnosis was ≥24,4 times the laboratory cut-point.

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AB0978 ASSOCIATION OF INFLAMMATORY ARTHRITIS WITH VOGT-KOYANAGI-HARADA SYNDROME (VKHS)

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Background: VKHS is a rare cause of granulomatous uveitis leading to significant visual loss in patients who develop it, usually accompanied with extraocular manifestations that include meningismus, vitiligo, poliosis, and hearing loss. In our country is responsible for 13% to 27% of all uveitis, affecting mainly young women.

Objectives: To describe the presence of inflammatory arthritis in patients with VKHS at a third level hospital.

Methods: A cross-sectional study of 4 patients with established VKHS that fulfilled the 2001 revised diagnostic criteria for Vogt-Koyanagi-Harada disease (1) was rolled out. Ultrasonography was performed to all patients by a trained rheumatologist in carpal, metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints evaluating presence of power Doppler (PD) signal, bone erosions, and cartilage changes.

Results: We included 3 women and 1 man, mean age was 34.7±10.3 years, and they all had characteristics of complete VKHS. Mean disease duration (since first manifestation of ocular symptoms) was 5.2±3.5 months. One patient had synovitis on physical examination. Out of the 4 patients with PD signal, 4 had involvement of both wrist, MCP and PIP joints consistent with polyarthritis pattern, and bone erosions were detected in one patient (Table)

Table. Characteristics of patients with VKHS and inflammatory arthritis

	Sex	Age	Complete VKHS	Serous retinal detachment	PD in carpal joints	Number of MCP joints with PD	Number of PIP joints with PD	Number of erosions	ESR/CRP elevation	Reumatoid Factor/ACCP
Patient 1	F	21	Yes	Yes	Yes	4	3	0	Yes	No
Patient 2	F	36	Yes	Yes	Yes	7	3	0	Yes	No
Patient 3	M	36	Yes	Yes	Yes	6	4	0	Yes	No
Patient 4	F	46	Yes	Yes	Yes	7	1	2	Yes	No

VKHS: Vogt-Koyanagi-Harada syndrome; PD: power Doppler signal; MCP: metacarpophalangeal joint; PIP: proximal interphalangeal joints, ESR: erythrocyte sedimentation rate; CRP: C reactive protein; ACCP: anti-cyclic citrullinated peptide

Conclusions: The presence of inflammatory arthritis in patients with VKHS has only been described in one case (2). Despite the exceptionality, we propose that polyarthritis and probably erosive arthritis can represent part of the spectrum of the disease, processes that share some features of the genetic susceptibility with rheumatoid arthritis as HLA-DR4, CTLA-4 and STAT4.

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AB0979 EOSINOPHILIC FASCITIS: CLINICAL EXPERIENCE IN A SERIES OF 21 PATIENTS

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Background: The Eosinophilic fascitis (EF) is a uncommon sclerodermiforme síndrome with unknown etiology and poorly understood pathogenesis and natural evolution. The published series are short, with a total of 280 cases described in the literature from 1974.

There is neither clinical tests nor consensus on the EF treatment.

Objectives: To describe a series of 21 patients with EF.

Methods: Multicentric, retrospective Case Series Study.

We reviewed 21 patients diagnosed of EF (cutaneous induration+consistent biopsy) between January 1998 to January 2015. A total of 13 males and 8 females.

Results: The mean age at diagnosis was 41 years 8range between 28 and 66).

Trigger factor was suspected in 8 patients (intense exercise in 7).

The zones of cutaneous affectation were fundamentally the upper and low extremities.

The hyperesonophilia was present in 100% of the patients. The average of the % more high of hyperesosinophilia was 21.2% (range 4–45.4%).

11 patients presented artralgias and 5 patients presented arthritis (4 polyarticular/1 oligoarticular).

The MRI was used for diagnosis and/or follow-up in 8 patients, especially in the diagnosed ones in the last 5 years.

The average initial dose of prednisone was 40 mg/día, the average maximum dose of prednisone was 40 mg/dia and the mean steroid treatment duration was 22 months (range 9–47 months). Other immunomodulatory agents were: methotrexate 11, hydroxychloroquine 2, azatioprina 2 and D penicillamine 1.

5 years after follow-up, the evolution was the complete resolution in 13 patients and the partial improvement in 6.

Other clinical and analytical manifestations (N=21).	YES, n (%)	NO, n (%)
Hypergammaglobulinemia	13 (63)	8 (37)
Elevated ESR	4 (16)	17 (84)
Elevated CRP	8 (37)	13 (63)
Elevated CK	0	21 (100)
Systemic symptoms	7 (32)	14 (68)
Visceral affectation	3 (11)	18 (89)
Autoimmune thyroid	2 (5)	19 (95)
Peripheral neuropathy	4 (16)	17 (84)

Conclusions: In our series, the EF prevails in males, in the decade of the 40 and in the majority one did not find trigger factor. There was presence of arthritis in a significant percentage of patients. In the last years the use of the RMN has been added to the diagnosis and to the follow-up. Though there are no clinical tests that support it the use of the metotrexato has been generalized as adjuvant treatment to the steroids.

Disclosure of Interest: None declared

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AB0980 EPIDEMIOLOGICAL PROFILE OF THE PULMONARY DISEASES IN MIXED CONNECTIVE TISSUE DISEASE: ABOUT 55 CASES

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Background: Mixed connective tissue disease (MCTD) are a systematic autoimmune disease that the aetiopathogeny remains misunderstood. This disease is often associated with systemic involvement which sometimes is very serious, such as pulmonary diseases.

Objectives: To describe the characteristics of the pulmonary diseases among a sample of MCTD.

Methods: We conducted a retrospective study including patient's cases diagnosed with a MCTD between the period of 10 years in the departement of internal medicine. The demographic, clinical, treated in the immunological, biological and radiological data were collected then analysed.

Results: In total we had 55 patients. 87% of them were female. The mean age of the sample was 50, 64±15 years. Major types of MCTD were: Rheumatoid polyarthritis (45.5%), sarcoidosis (18.2%), scleroderma (12.7%), and the lupus erythematosus (10. 9%). These causes were diagnosed after a mean delay of 686, 18 days. A lung disease was found in 50.9% of the cases. It was either present without symptoms in 52.8% of the cases or revealed by dyspnoea (22.6%), a cough (18.9%)or hemoptysie isolated or associated to thoracic pains (1.9% each).The most frequent type of respiratory diseases were adenopathies (26.9%). Pleurisy, fibrosis, and interstitial lung diseases were described among 11.5% of the cases. Bronchiectasis, Lung arterial high blood pressure, pleural nodules, bronchi dilatation were reported in 7.7%. In only 3.8% we had patients with cancer and emphysema. Chest radiographs were normal in 65.5% of the

cases but we discovered interstitial lung diseases (12.7%), pleurisy (7.3%), pleural nodule and mediastinal enlargement (3.6%) and pneumopathy (1.8%). The same appearences were described in the CT scann normal in 18.5% but in different proportions: adenopathies (25.9%), interstitial lung abnormalities (14.8%), fibrosis (11.1%), bronchiectasis (7,4%) and bronchi dilatation, pleurisy, nodule and mediastinal enlargement (3.7%). Respiratory functional exploration were normal in 69.2% obstructive lung disease and restrictive pulmonary disease were found within 11.5% of the patients.
Conclusions: Improvement of the knowledge of these diseases will improve the care before the appearance of complications.
Disclosure of Interest: None declared
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AB0981 CLINICAL FEATURES OF 28 CASES OF LIMB RESTRICTED VASCULITIS AND FASCIITIS

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Background: We sometimes experience the cases with fever and muscle pain of lower limbs without any other specific features. There are sporadic case reports of eosinophilic fasciitis and limb restricted vasculitis. However, few reports compare and discuss such cases.
Objectives: To describe the clinical features, MRI findings, histopathology, diagnosis and response to treatment of these cases.
Methods: We retrospectively analyzed the clinical features of 28 patients who were admitted to our hospital because of fever and muscle pain of lower limbs from 2004 to 2016.
Results: Among the 28 patients, 17 were vasculitis syndrome; eleven were limb restricted small vessel vasculitis (LrSvv), six were microscopic polyangiitis (MPA). Seven were fasciitis; three were eosinophilic fasciitis, three were diffuse fasciitis without eosinophilia and one was tuberculous fasciitis. One was relapsing polychondritis, one was Behçet's disease and the other two were myalgia without specific diagnosis. In our study, average age was 57.5±19.9 years old and older than in previously reported cases of limb ristricted vasculitis[1,2]. Sixteen were female, twelve were male. Abnormal MRI findings in non-infectious fasciitis and vasculitis syndrome were bilateral. Tuberculous fasciitis showed specifically abnormal intensity and fluid collection in unilateral thigh. Unilateral lesion and fluid collection may indicate infectious disease and bilateral lesion may indicate autoimmune or autoinflammatory diseases. MRI of vasculitis syndrome and fasciitis showed hyperintense T2-weighted signals in muscles of either legs, or thighs, or both. (n=3), MPA (n=1) and fasciitis (n=3). On MRI scan, abnormal fascial signal intensity was seen in all the patients with fasciitis and 6 (40%) with vasculitis syndrome. It was difficult to differentiate between vasculitis and fasciitis by MRI findings. Muscle biopsy was performed in 25 patients. In most cases, we performed en bloc biopsy, including muscle, fascia, skin and subcutaneous tissue. MRI was useful to determine the location of biopsy. All patients were treated with glucocorticoids. Immunosuppressive agents (azathioprine, n=10; methotrexate, n=5; cyclophosphamide, n=1; tacrolimus, n=1) were added in 15 patients and anti-tuberculous drugs in one. None of the 11 patients with LrSvv showed positive blood tests of anti-neutrophil cytoplasmic antibody or developed any other organ involvement during follow-up period (median 96 months; range 3–125). They responded well to glucocorticoid therapy (oral prednisolone 0.5–0.6mg/kg/day or intravenous methylprednisolone at doses of 1g/day). Recurrence rate of LrSvv patients was 0%, although that of MPA patients was 50% (n=3). In four patients with LrSvv, treatment was ceased and they achieved drug-free remission. There were no apparent differences between the patients who achieved drug-free remission and who didn't.
Conclusions: MRI and muscle biopsy were useful for diagnosis of disease with fever and muscle pain of lower limbs.
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AB0982 ANALYSIS OF THE TREATMENT EFFICACY FOR RETROPERITONEAL FIBROSIS - A CASE SERIES. SUSPICION OF ERDHEIM-CHESTER DISEASE IN A PATIENT WITH AN INITIAL DIAGNOSIS OF RETROPERITONEAL FIBROSIS, REFRACTORY TO STANDARD TREATMENT

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Background: Retroperitoneal fibrosis (RPF) is a rare disorder characterized by the development of fibrotic tissue in the retroperitoneum involving the abdominal aorta, iliac arteries and ureters. Erdheim-Chester disease (ECD) is a rare, non-Langerhans histiocytosis. Symptoms of ECD often resemble retroperitoneal fibrosis.
Objectives: Evaluation of standard treatment efficacy in 13 consecutive RPF patients. Revision of the diagnosis in patients unresponsive to standard therapy.
Methods: A retrospective analysis of 13 consecutive RPF patients (10 males and 3 females) treated in our department since 2008 was performed. All patients were treated with tapered dose of steroids combined with an immunosuppressive agent (cyclophosphamide in 4, azathioprine in 9, methotrexate in 6, hydroxychloroquine in 5 patients). Urologic interventions were undertaken as necessary.
Results: The treatment was effective in 10 patients: reduction of the retroperitoneal mass in computed tomography (CT) and normalization of the laboratory tests, including markers of inflammation and creatinine concentration. No improvement was found in 3 patients. However, in a further diagnostic work-up the diagnosis of RPF was maintained only in one of these 3 patients. In the second pateint an ANCA-asssotiated vasculitus was ultimately diagnosed. The third patient was treated with a combination of steroids and cyclophosphamide without any radiological improvement. However, the revision of CTs revealed the presence of changes typical of Erdheim-Chester disease.
Conclusions: The standard treatment (based on a combination of steroids and immunosuppressive drugs) is efficient in most partients with RPF. In refractory cases an alternative diagnosis, such as systemic vasculitis or Erdheim-Chester disease, should be taken into account.
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AB0983 MEFV MUTATIONS IN ARMENIAN PATIENTS WITH SYSTEMIC AUTOIMMUNE DISEASES

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Background: The systemic diseases of connective tissue have autoimmune mechanism in development. While autoimmunity involves adaptive immune activation, autoinflammation involves innate immune activation. The prototype of autoinflammatory diseases is familial Mediterranean fever (FMF), which is the global medical problem for Armenian ethnos on the whole, affecting 1–2% of population.
Objectives: The aim of this study was investigation of MEFV mutations and their possible influence on the systemic diseases in Armenian patients.
Methods: We have examined 183 patients with FMF. All patients with FMF fulfilled Tel-Hashomer FMF diagnostic criteria. Molecular-genetic detection of 12 MEFV mutations common for Armenians carried out in Medical Genetic Centre of Armenia. In 49 patients one of autoimmune systemic diseases was diagnosed: in 24 patients- seronegative spondyloarthropathy (SNSA), in 23 - systemic lupus erythematosus (SLE), 1 patient developed Sjogren's disease and 1- systemic sclerosis.
Results: In the SNSA group from 24 patients 16 were male, 8 were female, the mean age of patients was 35,4±12,2.The mean age at the beginning of the disease was 14,91±12,6. In all cases the symptoms of FMF were preceded symptoms of arthritis. Unilateral sacroiliitis was revealed in 6 patients, bilateral sacroiliitis-in 18 patients. The limitation of lumbar motion was assessed by Schober's test. 7 patients with Schober's test 1–2 cm had bilateral sacroiliits grade III-IV and fulfilled the modified New York criteria for ankylosing spondylitis. HLA B-27 was examined in 7 patients. In 5 cases it was negative, and in 2 cases –positive. MEFV gene analyses were carried out in 21 cases: 7 patients had one heterozygote mutations: 6-M694,1-M680I; 5 patients -M694V/M694V, 9 patients had compound heterozygote mutations: 5- M694V/V726A, 3 - M694V/E148Q, 1 - M680I/E148Q. So, the prevalent mutation was M694V.
In SLE group from 23 patients female were 21 (91.3%), male – 2 (8.7%). Mean age of patients was 37.4±2.5 years. The beginning of FMF was earlier than SLE. The activity of SLE estimated by SLEDAI index was significant lower than in SLE without FMF. SLE co-occurring with FMF had mild duration than classic lupus according to both clinical and laboratory findings including serological markers of SLE -ANA, anti-dsDNA. The prevalent mutation was M694V - 44.6%; V726A composed 21.7%, M680I-9.8%. Most common variations with M694V were followings:M694V/M694V,M694V/V726A,M694V/N.
Conclusions: A remarkable overlap was highlighted between FMF and SLE: both diseases have such common features as arthralgia, myalgia, arthritis, fever, skin involvement, serositis and renal involvement. It is likely, that the moderation in disease phenotype and peculiar disease characteristics observed in patients with both SLE and FMF are related to MEFV. MEFV mutations appear to modify SLE phenotype. Sacroilitis may be seen more frequently in FMF patients than expected. On the other hand, FMF must be kept in mind if patients not responding to the usual therapeutic interventions for sacroiliitis.
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