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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