with mesenteric panniculitis without skin and SCF involvement; the remaining 2 were not biopsied in hard to access areas. Histopathological study ruled out the probability of neoplasms and confirmed the diagnosis of lobular panniculitis in all specimens.

Therapy included such common in rheumatology practice agents as glucocorticosteroids, NSAIDs, cytotoxic drugs, hydroxychloroquine, and oths). Therapeutic success was documented in 62.68% cases, therapeutic failure and disease progression – in 17.91% (12 patients), requiring dose escalation and modification of therapeutic regimen.

Conclusions: Identified correlation between clinical features and lab parameters measuring disease activity confirms ILP as a systemic inflammatory disease of the connective tissue. There's a flagrant necessity to improve physicians awareness of ILP, as well as need in future studies to enable earlier ILP diagnosis and identify more effective treatment of the disease.

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AB1154

INCREASE GENERATION BUT DEFECTS OF SECRETING IFN- α PLAY A ROLE IN THE PATHOGENESIS OF IGG4-RD

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Background: IgG4 related disease (IgG4-RD) is a multi-organ involvement, fibro-inflammatory disease of unknown etiology. Both innate and adaptive immunity played vital roles in the pathogenesis of IgG4-RD. Plasmacytoid dendritic cells (pDC) had major roles in antigen presentation and secreting IFN- α upon infection. However, the characteristics and relevant function of this cell population in IgG4-RD was poorly understood. So we aim to study the expression and function of pDC in IgG4-RD.

Objectives: To study the expression and function of Plasmacytoid dendritic cells (pDCs) in IgG4-RD

Methods: Flow cytometry was performed to analyse the expression of pDC cells in untreated IgG4-RD patients (n=12) and healthy controls (n=12). The immunohistochemistry technique was used to assess the location of pDC in the involved tissues of IgG4-RD patients. Furthermore, by cells culture *in vitro*, the abilities of pDC secreting INF- α and the activation of NF- κ B signal in IgG4-RD were explored.

Results: The frequencies of pDC in the IgG4-RD patients were significantly higher in the peripheral blood and involved tissues compared with healthy controls. The cell surface marker of CCR7 in pDC was lower in untreated IgG4-RD patients, but after treatment, the expression of CCR7 increased. There was on significance of the expression of pDC maturation marker (CD83), activation marker (CD80, CD86), CCR1, CCR2, CCR3, CCR4, CCR5, CCR6, CCR8, CCR9, CCR10, CXCR2, CXCR3 and CX3CR1 compared with healthy controls. Interestingly, the frequencies of CD123hiCD303+cells were also higher in untreated patients, and reduced after treatment, and the cell surface marker CD83 was also elevated. By cell culture *in vitro*, pDC had defects in secreting INF- α of IgG4-RD patients than healthy controls.

Conclusions: The excessive infiltration of pDC in peripheral blood and tissue but less CCR7 Defects of secreting INF- α of pDC in IgG4-RD may indicate less function of eliminating infection which may induce constant infection. pDC may played vital roles in the pathogenesis of IgG4-RD,

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AB1155

IMMUNOGLOBULIN G4 – RELATED DISEASE, A DIAGNOSTIC CHALLENGE

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Background: The IgG4-related disease (IgG4-RD) is a chronic, inflammatory, multi-organic, systemic disease. The pathological assessment is the gold standard for the diagnosis, hallmarked by the lymphoplasmacytic tissue infiltration of mainly IgG4-plasma cells, the storiform fibrosis and the obliterative phlebitis.

Objectives: The aim of this study was to analyse the features of IgG4- RD cases followed in a specific autoimmune diseases unit, since 2013.

Methods: Descriptive, retrospective study, through the review of clinical charts. Medical records were reviewed for demographics information, clinical

presentation, underlying conditions, laboratory and radiological data, medical and surgical treatments and clinical outcomes. All patients diagnosed by biopsy were included (international consensus pathological criteria of 2012). We excluded the possible cases of IgG4-RD when biopsy was not available or did not meet the criteria previously mentioned.

Results: Eight patients with characteristic clinical, histological and laboratory features of IgG4-RD were included. The patients were predominantly male (87.5%), the mean age at diagnosis was 66±11 years.^{48–80} 87.5% of the cases were consulted from other hospital Services, mainly surgical: Urology 25% (hydrocele and urinary retention), General Surgery 25% (obstructive jaundice and forearm tumour), Vascular Surgery 25% (abdominal aortic aneurysm), Cardiac Surgery 12.5% (thoracic aortic aneurysm) and Neurology 12.5% (hypertrophic pachymeningitis). The most commonly involved organs were vascular 87.5% (6 aorta, 1 pulmonary veins) and retroperitoneum 75%, followed by renal 37.5%, pancreatic 12.5%, and central nervous system 12.5%. It was found isolated organic involvement in only one patient (autoimmune pancreatitis). Two patients had previous malignancy: renal cells and prostate. Magnetic resonance imaging, computed tomography and FDG-PET were made in 62.5% each of them. Serum IgG4 was determined in all cases, 62.5% of patients had normal IgG4 serum levels (the upper limit of normal for serum IgG4 is 135 mg/dL). Three patients (37.5%) had elevated serum IgG4, the mean level was 188±38 mg/dL ((152–266). The pathological findings were dense lymphoplasmacytic infiltrate in 6 cases (75%) with obliterative phlebitis in 5 of them, storiform-type fibrosis in 4 cases (50%) and IgG4/IgG ratio >40% in 75% of the samples. Glucocorticoids treatment was initiated in 75% of patients, 25% were untreated for predominantly fibrotic involvement. There were treatment response in 60% of cases. 25% had a relapse after corticoid withdrawal. Only one patient received a second drug due to lack of response to Prednisone (rituximab).

Conclusions: According to the literature, IgG4-RD patients were predominantly male in their sixth and seventh decade of life. The predominant involvement in our serie was vascular and retroperitoneal, with no cases of glandular manifestations. Highlight the high number of cases with normal serum IgG4. Most of our patients responded to corticosteroid therapy.

Disclosure of Interest: None declared

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AB1156

DRESS (DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS): LOOKS MAY BE DECEPTIVE!

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Background: Drug reaction with eosinophilia and systemic symptoms (DRESS) is a rare, potentially life threatening multisystem disorder with fever, skin eruptions, lymphadenopathy, eosinophilia and systemic involvement most commonly after a drug exposure. These cases may mimic various rheumatological conditions. We report a series of 14 cases of DRESS who presented to Rheumatology Clinic as suspected connective tissue disease or sepsis in Northern India.

Objectives: To highlight DRESS as a common mimic of common rheumatologic conditions and sepsis and report its etiology, characteristics, treatment and prognosis

Methods: We manually searched the inpatient records of Immunology Department in SGPGI 2007–18 for the cases discharged with a diagnosis of possible/probable/definite DRESS. The records of the patients with probable and definite DRESS according to regiSCAR criteria¹ were reviewed.

Results: All the 14 patients fulfilled clinical criteria for diagnosis (4 probable, 10 definite). The age of patients ranged from 9–53 years with majority in their 3rd decade. Majority of the patients were referred to us when their counts were rising in the setting of fever and skin rashes with a suspicion of rheumatic disease/sepsis. The clinical and laboratory features of these patients are as follows (table 1): TLC: Total Leucocyte Count, AEC: Absolute Eosinophil Count, ED: Exfoliative dermatitis, F: facial oedema, M: maculopapular rash, L: Liver, K: Kidney, P: Lung, G: GI, SSZ: Sulphasalazine, HRZE: Isoniazid, Rifampicin, Pyrazinamide, Ethambutol, FU: Follow up

All the patients were treated with oral steroids showing signs of clinical improvement within 4–5 days. Rashes and leucocyte count were first to respond. Transaminitis responded within a week. Mean Followup is 9.3 months. They were gradually tapered off steroids over next 3–4 months except for two patient who were lost to follow up.