Sarcoidosis (SA) a multiorgan granulomatous disease. The inflammatory process in involved organs is characterized by Th1/Th2 imbalance and participation of Th17 cells, what was recently documented. SA is more prevalent among non-smokers (NS)). The concentrations of IL-17A, IFN-γ, TNF-α, IL-10, IL-6, IL-4 and lower of IL-2 in S than in NS (1.28 ± 0.05 vs 1.23 ± 0.05, r=0.3, P<0.05, respectively). The mean concentration of IL-17A was 6.1 ± 0.2 pg/mL and did not differ between S and NS. There was significant correlation of INF-γ with the extension of fibrosis.

Conclusions: This study validates a novel risk score for prediction of outcome in patients with idiopathic pulmonary fibrosis.

References:

P3613
Expression of interleukin-27 (IL-27) in human lower airways.
Pathophysiological implications in pulmonary sarcoidosis (PS)
Piotr Kopinski1, Tomasz Wnuk1, Joanna Chorostowska-Wynimkó1, Grzegorz Przybylski1, Ewelina Polęgsek1, Andrzej Dyczek2.
1Dept. of Gene Therapy, Collegium Medicum, Nicolaus Copernicus University, Bydgoszcz; 2Laboratory of Molecular Diagnostics and Immunology, Institute of Tuberculosis and Lung Diseases, Warsaw; 3Dept. of Lung Diseases and Tuberculosis, Collegium Medicum Nicolaus Copernicus University, Bydgoszcz, Poland

Background: Interleukin-27 was recently discovered as a cytokine secreted by antigen presenting cells, participating in T cell priming and supporting Th1 type immune polarization. IL-27 expression has not been examined yet in lower airways.


Methods: Intracellular IL-27 expression cells was determined by indirect phenotyping and flow cytometry in BAL cells obtained from patients with PS (incl. steroid treated subjects), idiopathic pulmonary fibrosis (IPF) and controls, as well as in Human Lung Fibroblasts (HLF1) and pneumocyte type II (A549) cell lines. IL-27 extracellular secretion was tested by ELISA (cat. no E90385Ha, Diaclone) in supernatants of BAL and cell cultures.

Results: BAL detectable IL-27 levels were shown by ELISA in 6 (median 9 ng/ml) of 13 PS untreated patients. IL-27 was not found in BAL supernatants of steroid-treated PS, IPF and controls. HLF1, A549 and alveolar macrophages were positive for intracellular IL-27. Surprisingly, IL-27+ BAL lymphocytes were found in all tested groups (PS: 81±15%, median 30%, IPF: 37±15%, median 23%).

Conclusions: This study validates a novel risk score for prediction of outcome in patients with idiopathic pulmonary fibrosis.
Conclusions: In physiological conditions, IL27 is produced in lower airways by lung fibroblasts, epithelial and BAL immune cells, including lymphocytes. Its increased expression in PS suggests IL27 to play a role in ILD pathophysiology, probably as Th1 activity marker.

P3614 Pulmonary graft-versus-host disease (GVHD) post- stem cell transplant (SCT) for haematological malignancies: Good response to treatment if diagnosed early
Bhargavshree Jayaraman1, F. Begum2, K. Raj1, H. Milburn1. 1Department of Respiratory Medicine, Guys Hospital, Guys & St Thomas’ NHS Trust, London, United Kingdom; 2Medicine, Kings College, London, United Kingdom

Introduction: Graft-Versus-Host Disease (GVHD) of the skin and gut are relatively common following allogeneic transplant for haematological malignancies but GVHD affecting the lung can be subtle.

In this study, we evaluated patients who developed lung GVHD post stem cell transplant (SCT) to determine early warning signs and response to treatment.

Method: Patients undergoing SCT for haematological malignancies were monitored with serial pulmonary function (PFT). Symptoms of increased breathlessness (SOB) or fall in PFT prompted investigation with high resolution CT scan (HRCT).

Results: 11 patients developed lung GVHD following allogeneic SCT for lymphoma (n=5) or leukemia (n=6). 6 had adjuvant chemotherapy, and 7 total body irradiation. 10 had exertional SOB. 3 developed pulmonary GVHD within 1 year of SCT.

PFT changes are shown.

All patients had mosaicism with gas trapping on HRCT at diagnosis, and were commenced on steroids, with 6 having other immunosuppressants.

The post-treatment HRCT showed clearance in 10 patients (1 died due to relapse of malignancy).

Conclusion: Exertional SOB and changes in PFT are important early indicators of developing GVHD which can be confirmed on HRCT. Treatment with immunosuppressants results in successful resolution in the majority, particularly in those who were diagnosed early in the course of the complication.

P3615 Response to physical exercise in sarcoidosis
Katarzyna Hildebrand

Exertional SOB and changes in PFT are important early indicators of developing GVHD which can be confirmed on HRCT. Treatment with immunosuppressants results in successful resolution in the majority, particularly in those who were diagnosed early in the course of the complication.

Conclusion: Exertional SOB and changes in PFT are important early indicators of developing GVHD which can be confirmed on HRCT. Treatment with immunosuppressants results in successful resolution in the majority, particularly in those who were diagnosed early in the course of the complication.

P3616 Isolated lymphocytic bronchiolitis with B cell clonality: Diagnosis in 3 cases using immunoglobulin gene rearrangement analysis
Irina Lings1, Vincent Condon1, Françoise Thivolet-Bejot2, François Berger3, Chahéta Khouastra1, Baakar El Zoubi1, Gilles Salles2, Jean-François Cordier1.
1Respiratory Medicine - Reference Center for orphan Pulmonary Disease, Louis Pradel University City Hospital, Lyon, France; 2Hematology, CH Lyon Sud, Lyon, France; 3Pathology, CH Lyon Sud, Lyon, France; 4Pathology, Louis Pradel University City Hospital, Lyon, France

Background: Lymphocytic bronchiolitis is characterised by diffuse infiltration of the bronchiolar walls by lymphocytes, which may organise into germinal centres (follicular bronchiolitis). B cell clonality demonstration may result in a diagnosis of low-grade B-cell lymphoma.

Patients and methods: Patient #1: 59-yr old female, non-smoker, presented with chronic cough, dyspnea, sicca syndrome, negative anti-SSA/SSB antibodies, severe airflow obstruction, ground glass opacities at HRCT, and 51% of lymphocytes at BAL. Patient #2: 54-yr old female, non-smoker, presented with chronic cough, restrictive lung disease, diffuse micronodules at HRCT, 90% lymphocytes at BAL, monoclonal IgG lambda of 4.2 g/l. Patient #3: 66-yr old female, non-smoker, with history of rheumatoid arthritis and Sjogren syndrome, anti-SSA and -SSB antibodies, presented with hemoptysis, Bence-Jones protein, airflow obstruction, 2 lung amyloid nodules and multiple cystic lung disease.

Results: Lung biopsy with immunohistochemistry demonstrated lymphocytic bronchiolitis in all three cases with a majority of CD 20 positive B- cells. PCR-based DNA testing for immunoglobulin gene rearrangement analysis of lung biopsy demonstrated B-cell clonality in all three cases (VK-JK, FR1-JH), and low-grade MALT lymphoma was eventually diagnosed.

Conclusion: Lymphocytic bronchiolitis may correspond to low-grade MALT lymphoma which may be demonstrated by gene rearrangements rearrangement analysis.

P3617 The relationship between leptin and proinflammatory cytokines with bone mineral density in patients with sarcoidosis (pts)
Elżbieta Puceńska1, Michal Bednarek1, Anna Gelgaj-Geremek1, Anna Kowalczyk2, Joanna Chorostowska-Wynimko2, Paulina Jagus2, Ada Roży2, Jacke Grudny1, Dariusz Chmielewski3, Dorota Gorecka4. 1Department of Respiratory Medicine, National Institute of Tuberculosis and Lung Diseases, Warsaw, Poland; 2Laboratory of Molecular Diagnostics and Immunology, National Institute of Tuberculosis and Lung Diseases, Warsaw, Poland; 3Department of Lung Diseases, National Institute of Tuberculosis and Lung Diseases, Warsaw, Poland; 4Department of Orthopaedics and Traumatology of Locomotor System, Medical University of Warsaw, Poland

Background: Sarcoidosis is a chronic inflammatory disease. Osteoporosis is a multifactorial disorder of reduced bone mass. Body weight is commonly considered a significant predictor of bone mineral density (BMD) but obesity is also associated with chronic inflammation. Cytokines like TNF-α, IL-1 and IL-6 induce bone resorption and bone loss.

Aim: The aim of the study was to investigate whether there is a relationship between leptin or cytokines and BMD in pts.

Methods: 85 pts, none was treated with steroids, 46M and 39F, age 41±9y were enrolled. We evaluated BMI, serum leptin, TNF-α, IL-1 and IL-6, osteocalcin, alkaline phosphate and others. BMD was measured (DXA).

Results: Pts with low BMI had decreased lumbar spine BMD (Tscore<−1). The differences were statistically significant for group with normal BMI(18,5<−BMI<−25), compared to overweight (BMI<−30), p=0,045 and to obese (BMI>−30), p=0,0054. There were no such results for femoral neck BMD. The highest values of leptin were found in pts with obesity. The differences between pts with normal BMI compared to those with obesity was statistically significant (p=0,026). There was a weak correlation between leptin and BMI, R=0,25, leptin and IL-1,R=0,27, leptin and phosphate in urine, collected over 24h, R=0,31. No correlation was found between leptin and TNF-α or IL-6.

Conclusions: In our group BMI was related to BMD but not to leptin levels nor other cytokines. Obesity, through its mechanical loading effect have a protective influence on bone tissue metabolism. Leptin, cytokines have a role in bone growth, development and loss. Further research is required to ascertain the importance of adipokines or cytokines for BMD.

P3618 Multinodular parenchymal pulmonary amyloidosis in primary Sjögren’s syndrome: A case report
Elena Soma Ozyilmaz1, Ozlem Cancoz2, Fatih Kurnaz2, Mustafa Çalis1.
1Afa Yildirim1, 2Non Tutar1, Ayse Kanbak1, Hakun Buyukkag1, Inci Gelmez1, Runanaz Demir1. 1Department of Chest Diseases, Erciyes University Medical School, Kayseri, Turkey; 2Department of Pathology, Erciyes University Medical School, Kayseri, Turkey; 3Department of Internal Diseases, Hematology Unit, Erciyes University Medical School, Kayseri, Turkey; 4Department of Rheumatology, Erciyes University Medical School, Kayseri, Turkey; 5Department of Physical Therapy and Rehabilitation, Erciyes University Medical School, Kayseri, Turkey; 6Department of Radiology, Erciyes University Medical School, Kayseri, Turkey

Amyloidosis is a rare cause of pulmonary infiltrates in primary Sjögren’s syn-
P3619 Sarcomiosis is a frequent benign cause of lymphadenopathy in neoplastic patients
Claudia Ravaglia, Carlo Gurioli, GianLuca Casoni, Micaela Romagnoli, Sara Tomassetti, Christian Gurioli, Poletti Venerino.
Department of Pulmonology, Forlì, FC, Italy

Background: Clinical and radiographic aspects of sarcomiosis and malignancy might mimic one another, making the distinction between the two difficult in some cases. Cancer and sarcomiosis have been described in some case series from the literature but this association remains controversial. Objectives: A descriptive retrospective study to evaluate the incidence of sarcomiosis in patients followed-up for previous cancer and referred for hilar/mediastinal lymphadenopathies, with no pulmonary lesions.

Methods: We conducted a retrospective chart review of all patients who were referred to our pulmonology department in the period between January 2007 and October 2011 with a new onset hilar/mediastinal lymphadenopathies during follow-up for previous neoplasms.

Results: Forty-eight patients (31 males, 17 females) of mean age 63 years (range 20-81) underwent EBUS/EUS trans-bronchial needle aspiration (TBNA). Patients had a diagnosis of lung cancer (10 cases), breast cancer (7 cases), urologic tumors (6 cases), gastric/intestinal lesions (6 cases), sarcomiosis (4 cases), and lympho(pharynx tumors (4 cases), gynecologic tumors (3 cases), other cancer (8 cases).

Conclusion: Sarcomiosis should be considered in the differential diagnosis of patients with a history of malignancy who develop hilar/mediastinal lymphadenopathies; a tissue diagnosis should be obtained before instituting therapy for presumed cancer recurrence.

P3620 Clinical study of pulmonary hypertension complicating pulmonary Langerhans' cell histiocytosis
Enna Li1, Haiqing Dai1, Luying Peng1, Mulan Jin2, Chen Wang1,1
1Department of Respiratory and Critical Medicine Care, Beijing Chao-Yang Hospital, Beijing, China; 2Department of Pathology Medicine, Beijing Chao-Yang Hospital, Beijing, China; 3Beijing Hospital, Health Ministry, Beijing, China

Objective: To study the clinical features of pulmonary hypertension (PH) complicating pulmonary Langerhans' cell histiocytosis (PLCH).

Methods: Medical records of 11 PLCH patients were reviewed from June 2006 to June 2011.

Results: 4 of the 11 PLCH patients presented PH with more severe clinical presentations. The major symptoms were laboring dyspnea and diminished exercise capacity (NYHA functional class IIIIV), which were complicated with the sight of right heart failure. The cystic change and pulmonary artery/main artery >1 were predominated on chest high-resolution computed tomography. Right heart enlargement was also found. As to pulmonary function presentation, the patients displayed severe carbon monoxydose diffusioning capacity impairment and significant hypoxemia. The pulmonary artery systolic pressure were highly increased. Besides the clinical symptoms, some features presented, involvement of both arteries and veins was also observed. Oxygen and sympathetic therapy was essential in the part of clinical treatment. 3 of the 4 PH-PLCH received corticosteroids or chemotherapeutic agents, but the effect was not ideal. Only one case of PH-PLCH had a stable condition.

Conclusion: Pulmonary hypertension is a common complication of PLCH and seems to predict a poor prognosis. It is necessary to derive an early diagnosis and prevention.
Thematic Poster Session

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P3624

A newly developed sling incorporating a shock absorber to minimize the motion effect on SpO2 during the 6-minutes-walk test
Kensu Miyamoto, Yuya Tanaka, Toshiaki Kurimoto. Department of Rehabilitation, Faculty of Health Sciences, Hokkaido University, Sapporo, Japan

Recently, the degree of desaturation during 6-minute-walk test (6MWT) is considered to have prognostic value in patients with idiopathic pulmonary fibrosis. In this study, we demonstrate a negative effect of motion on SpO2 during the walking and present a newly developed device that minimizes the negative effect during the test.

Subjects and methods: Ten healthy volunteers (26.1±11.7ys) performed 6MWT in 4 different manners: 1) usual walking, 2) usual walking while trying to minimize the effect of motion on the finger on which the oximeter-probe was placed, 3) usual walking using a newly developed sling with a shock absorber (75g) wrapped around the finger and the probe, and 4) walking on a treadmill at the same speed as the usual 6MWT while the finger and the probe were in a resting position without movement. SpO2 was measured continuously using a Pulsox 300i (Konica-Minolta).

Results: During the usual walking with/without the finger moving, there was severe desaturation of more than 4% from the baseline. However, when walking with the new device or on the treadmill there was only a small change in the SpO2, which was within the accuracy of the oximeter.

Conclusion: SpO2 monitoring during 6MWT does not provide reliable data on desaturation. However, using the new device, more reliable data on desaturation are available.

P3625

Effects of lymphadenopathy on pulmonary function tests in sarcoidosis
Omer Araz1, Elif Yilmazel Ucar1, Yener Aydin1, Melmet Meral1, Yusuf Bilen1, Fatih Alper1, Ali Metin Gorgenc1, Metin Akgan1, 1Chest Department, Ataturk University, Erzurum, Turkey; 2Thoracic Surgery, Atatürk University, Erzurum, Turkey; 3Radiology, Atatürk University, Erzurum, Turkey; 4Respiratory Medicine, Hôpitaux Universitaires, Paris-Seine-Saint-Denis (Avicenne), Paris, France

Background and objectives: The pulmonary function test (PFT) is an important diagnostic test in the follow-up of sarcoidosis. We aimed to investigate the effect of the presence of lymphadenopathy (LAP) alone or after controlling parenchymal involvement in PFT.

Materials and methods: A total of 63 sarcoidosis patients were enrolled retrospectively in the study. Respiratory function tests were evaluated via PFT. Radiological evaluations of the patients were done with chest x-ray and high-resolution computerized tomography (HRCT). Bronchoscopic investigations were performed on all patients.

Results: There is statistically significant correlation between the bronchoscopic findings and PFT parameters (p<0.01). Forced vital capacity (FVC) was affected more in the presence of LAP in bronchi neighborhoods, it was more or less the same for forced expiratory volume in 1 second (FEV1). Considering grade of HRCT findings, there is a statistically significant relation between FVC, FEV1, and the presence of hilar LAP, intrathoracic LAP and lobel LAP (p<0.001). There is a statistically significant correlation with the presence of micronodules in HRCT and all parameters of PFT. There is a statistically significant negative correlation between the radiological stage of sarcoidosis and FVC, FEV1.

Conclusion: We revealed that besides parenchymal involvement of the disease, special localization of lymph node involvement also has an important effect on the PFT parameters of sarcoidosis patients.
differentiation and migration of primary normal or fibrotic human lung fibroblast
(HLFs) in vitro.

Methods: Fibroblasts were derived from lung tissue of idiopathic pulmonary
fibrosis (IPF) patients and patients who suffered from primary spontaneous pneu-
mothorax. Real time RT PCR was used for the assay of mRNA level of profibrotic
cytokines in fibroblasts. Wound healing and migration assay was used for the
mobility analysis of fibroblasts.

Results: We demonstrated that Rapa strongly amplifies the increase in basal or
TGF-β1-induced fibronectin (FN), connective tissue growth factor (CTGF), TGF-
β1, α-smooth muscle actin (SMA), tissue inhibitor of matrix metalloproteinase
(TIMP)-1/plasminogen activator inhibitor (PAI-1) mRNA levels in normal or fi-
brotic HLFs. These positive functions of Rapa are in a Smad-independent way.

Wound healing and migration assay showed that Rapa could not affect the mobility
of HLFs.

Conclusions: These experiments demonstrate that Rapa positively regulates vari-
ous gene expressions of HLFs. We suggest another evidence that Rapa may exert
a profibrotic effects in vitro.

P3629
The markers of inflammatory process activity and fibrogenesis activity in
patients with idiopathic pulmonary fibrosis

O.V. Karas'ova, E.S. Khmel, E.N. Kovalenko, V.V. Rodionova. Hospital therapy
and Profpathology, Dnepropetrovsk Medical Academy, Dnepropetrovsk, Ukraine

Purpose: To define the inflammatory markers and severity of idiopathic pulmonary
fibrosis (IPF).

Design: There were examined 29 patients with IPF (men - 21, women - 8) in the
age of 35 - 57 years. Duration of disease was from 2-3 months till 2-3 years.
Control group included 10 practically healthy persons (donors).

Methods: Common clinical methods, X-ray examination, definition of hemostasis
parameters. The hardphase method of immune analysis was used for measuring
level of plasma fibronectin (PFN) and lavage fibronect (LFN). Patients had been
examined during the period of disease aggravation.

Results: Patients with IPF had increased level of PFN and decreased activated blood
recalcification time (ABRT) (p<0.05). The level of PFN was 319.2±14.7 mkg/ml, LFN - 0.055±0.011 mkg/ml and didn’t depend on both sex and age.

There existed straight connection between level of PFN and ABRT (r = 0.4353;
p<0.05) and reverse connections between levels of PFN and LFN and level of
plasma fibrin (r =-0.5482 and r=-0.4969; p<0.05). Severity of patients condition
correlated with increased level of PFN.

Conclusions: Revealed inflammatory markers and changes in hemostasis sys-
tem testify about activation of fibrogenesis and forming of chronic disseminated
intravascular coagulation.