

TUESDAY, SEPTEMBER 4TH 2012

radiograph showed patchy infiltration and consolidation near his left hilar. Chest CT scan showed patchy infiltration and round-shape, hypodense homogeneous lesions located in the lower lobe of right lung without any calcification. After 19 days of antibiotics treatment, his clinic symptoms and signs disappeared, he underwent chest CT scan (Figure 1) again after treatment, and the result showed the patchy infiltration dissolved, however, the lesions in the lower lobe of right lung remained without any change.

He was discharged. One month and one year later he was examined by chest MRI. The lesions were diagnosed as intrapulmonary lipoma by T2 weighted MRI images (Figure 2) with fat suppression.

Conclusions: Intrapulmonary lipoma can also be identified in children. Chest CT scan and MRI are very important for making correct diagnosis.

P3500

Munchausen syndrome by proxy presenting as unexplained hemoptysis:

A report of two cases

Suleyman Tolga Yavuz¹, Guzin Cinel², Ugur Ozelik², Ebru Yalcin², Deniz Dogru², Nural Kiper². ¹Department of Pediatrics, Pediatric Asthma and Allergy Unit, Hacettepe University, Ankara, Turkey; ²Department of Pediatrics, Pediatric Chest Diseases Unit, Hacettepe University, Ankara, Turkey

Hemoptysis is a serious symptom in early childhood and further investigation may be required in some cases. Munchausen syndrome by Proxy (MSBP) must be kept in mind particularly when suspicious behaviors of the supervisors are observed.

Case 1: A thirteen-month-old boy was admitted with recurrent hemoptysis. He was hospitalized because of persisting symptoms for four months, however no reason was found and he was subsequently discharged. Detailed history revealed that hemoptysis had occurred only in times when he was alone with his mother. Despite comprehensive diagnostic evaluation including laryngoscopy, Computerized Tomography, bronchoscopy and endoscopy, no definitive etiology was diagnosed. During his hospitalization, the mother reported new bleedings of the child and blood stained clothes were noticed. Examination of security footage revealed that the mother was sneakily obtaining the blood samples of another patient and deceiving medical staff.

Case 2: An eight-month old girl was admitted with a history of having vomited blood for two months. Reluctance of her mother was noticed by the physician during history taking. Upper endoscopic investigation revealed linear hemorrhagic lesions at pharynx and tonsils. No definitive etiology was diagnosed despite comprehensive investigations. During hospitalization, the doctor witnessed that her mother was injuring the child with her finger and making her bleed. She was warned about security footage of the room and her complaints resolved spontaneously.

Munchausen syndrome by proxy is an unusual and difficult to identify cause of child abuse. It must be taken into consideration particularly in conditions with secondary gain of family members.

P3501

Thoracic splenosis – Case series and review of the literature

Nissim Arish¹, Ariel Rockach¹, Naama Bogot², Irith Hadas-Halperen², Gabriel Izibicki¹. ¹Pulmonology, Shaare-Zedek Medical Center, Jerusalem, Israel

Introduction: Splenosis is a rare finding of ectopic splenic tissue found within the thoracic cavity, abdomen or peritoneal cavity. In thoracic splenosis the splenic tissue most often grows in the form of a nodule and the auto-transplantation is usually caused by a previous penetrating or blunt trauma to the thoracic-abdominal region, resulting in splenic rupture.

Cases: We present three cases of thoracic splenosis. All the patients were Caucasians male aged 40-63 presenting with pleural masses that mimicked malignancy, who were eventually diagnosed as thoracic splenosis. All of them had a history of traumatic event to their upper abdomen which occurred 13-40 years ago resulting in splenectomy.

Methods: We made a Medline search in order to find all the cases of thoracic splenosis. We reviewed the cases in order to specify this rare condition.

Results: We found 39 case reports of patients diagnosed with thoracic splenosis. In the majority of the cases the patients are asymptomatic and are incidentally diagnosed with left hemithorax pulmonary lesions found via chest radiography or thoracic computed tomography. The average period of diagnosing thoracic splenosis from the time of trauma is roughly 21 years and ranges from 6 to 46 years. The diagnosis is challenging and the patients usually undergo extensive workout including surgical biopsy

Conclusion: We conclude that splenosis is a rare entity but one should think about this possibility in a patient with a left thoracic mass and a history of abdominal trauma, splenic rupture and splenectomy.

Abstract P3502 – Table 1. Diagnosis, location, origin of arterial vessel, and symptoms

| Patient | Diagnosis | Location | Origin of Vessel | Follow up (yrs) | Exacerbations | Life threatening events |
|---------|--|----------|------------------------------|-----------------|---------------|-------------------------|
| 1 | MDCTA, MRA, Gallium Scan, CT guided Biopsy | LLL | TA | 11 | 0 | None |
| 2 | MDCTA | RLL | TA | 5 | 0 | None |
| 3 | MDCTA, PET-CT. | LLL | Abdominal Aorta/Celiac trunk | 6 | 0 | None |
| 4 | MDCTA, PET- CT. | RLL | TA | 11 | 0 | None |
| 5 | MDCTA | LLL | TA and Celiac Trunk | 6 | 2 | None |

CTA, computer tomography angiography; PET, Positron Emission Tomography; LLL, Left lower lobe; RLL, Right lower lobe; TA, Thoracic Aorta.

P3502

Pulmonary sequestration in asymptomatic adults; a plea for conservative observation

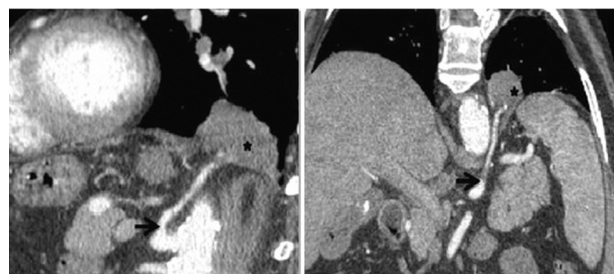
Artemios Dimitriou, Yehudit Rozenman, Issahar Ben Dov. *Pulmonary Department, Sheba Medical Center, Tel Aviv, Israel* *Radiology Department, Sheba Medical Center, Tel Aviv, Israel* *Pulmonary Department, Sheba Medical Center, Tel Aviv, Israel*

Background: Pulmonary sequestration (PS) is a rare congenital anomaly. Due to recurrent infections, the treatment in children is surgical removal of the sequestered lung and this recommendation expands to adulthood.

Objective: To test if asymptomatic or mildly symptomatic adults with PS could be offered conservative follow up instead of surgery.

Methods: We conducted a case series analysis of all patients above 20 yrs old, with sequestration seen in our institute from 2000-2012. Diagnosis, symptoms, patient characteristics, natural course and treatment were reviewed.

Results: During a median follow up of 7.8yrs, no major infection occurred and the lesion size remained unchanged in 4/5 patients while mild infection that resolved with oral antibiotics was noted in 1/5.



Conclusion: In asymptomatic or mildly symptomatic adult patient with lung sequestration, conservative follow up instead of surgery is a legitimate option.

P3503

Acquired tracheoesophageal fistula – A rare cause of chronic episodic cough

Amitesh Gupta¹, Saurabh Kansal², A.P. Kansal¹, Bidhi Chand¹. ¹Chest and Tuberculosis, Govt. Medical College, Patiala, Punjab, India; ²ENT, Dayanand Medical College, Ludhiana, Punjab, India

Introduction: Acquired tracheoesophageal fistula (TOF) is a rare entity and occurs as a result of trauma, malignancy, granulomatous infection, any previous surgery of trachea and oesophagus. Symptoms include uncontrolled coughing after swallowing. The majority of acquired fistulas occur at cervico-thoracic junction.

Case report: We report a case of 55 yr female who presented to us with cough and regurgitation particularly after intake of liquids since two mths. There was no history of tuberculosis or malignancy. Patient was on antihypertensive agent for four mths but not on ACE inhibitors. On routine investigations TLC and ESR were raised but chest xray was normal.

Sputum was negative for AFB. Patient was managed on lines of Gastro Esophageal Reflux Disease with no improvement. CECT chest showed consolidative changes. Nothing significant found in upper GI endoscopy. Barium swallow revealed the passing of contrast into trachea and in bronchi, showing the presence of tiny fistula at C3-C4 vertebral level. Due to higher position of fistula, patient was advised to undergo surgery.

Discussion: An acquired TOF bypasses the laryngeal protection and leads to repeated aspiration. We are reporting a rare case of acquired TOF possibly of infective etiology which is a rare occurrence at such a age. It was also the cause of chronic episodic cough in this patient which was not relieved with various treatments. Malignant and traumatic causes have now superseded infection. Barium swallow will demonstrate the defect in 70% of lesions. The management of acquired TOF involves minimizing further aspiration to prevent pulmonary infections. Definitive management involves stenting or surgical repair with treatment of underlying cause.

TUESDAY, SEPTEMBER 4TH 2012

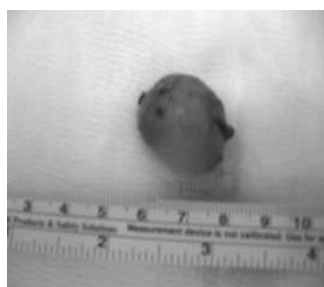
P3504**An unusual cyst of posterior mediastinum: Mullerian cyst**

Mehmet Dakak¹, Baris M. Poyraz², Safak Bulut³, Cihangir Dogu⁴, Selçuk Bilgin⁴, ¹Thoracic Surgery Department, Private TOBB ETU Hospital, Ankara, Turkey; ²Chest Diseases Department, Private TOBB ETU Hospital, Ankara, Turkey; ³Pathology Department, Private TOBB ETU Hospital, Ankara, Turkey; ⁴Intensive Care Unit, Private TOBB ETU Hospital, Ankara, Turkey

In this case, we report a newly categorized mediastinal cyst called Mullerian cyst, which was reported for the first time by Hattori et al in 2005. A 51-year-old female patient was admitted to our hospital with the complaint of dysphagia. T2- weighted chest magnetic resonance imaging (MRI) showed a cystic lesion attached to the vertebral column in the posterior mediastinum.



The patient underwent right thoracotomy. The cyst was tightly attached to esophagus and corpus of the 5th thoracic vertebra. The intact cyst was freed from posterior mediastinum.



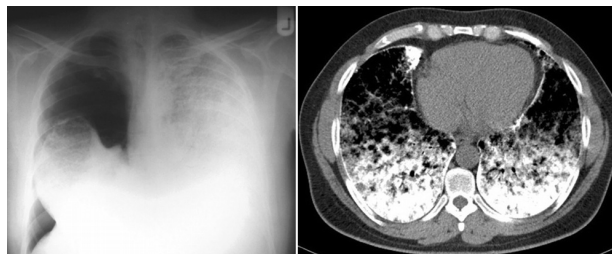
It was lined by a ciliated tubal-type epithelium and had a thin wall composed of smooth muscle. The epithelium showed estrogen and progesterone receptor expressions immunohistochemically. Mullerian cysts arise from remnants of the Mullerian duct and can be located anywhere along the path of Mullerian duct regression. Pelvis is the most common localization and their occurrence in the mediastinum is extremely rare. However mediastinal mullerian cyst is newly known entity, it should be kept in mind for the differential diagnosis of posterior mediastinal cysts.

P3505**First presentation of a case of pulmonary alveolar microlithiasis with spontaneous pneumothorax**

Naseh Sigari¹, Shilan Mohammadi², ¹Internal Medicine, Kurdistan University of Medical Sciences, Sanandaj, Kurdistan, Islamic Republic of Iran; ²Pediatric Medicine, Kurdistan University of Medical Sciences, Sanandaj, Kurdistan, Islamic Republic of Iran

Pulmonary Alveolar Microlithiasis (PAM) is a rare disease of unknown etiology. It is characterized by the presence of small calculi within the alveolar space. Clinical features vary and some patients may be asymptomatic for a long time with subsequent occurrence of dyspnea, dry cough, chest pain, and ultimately, respiratory failure. Recurrent spontaneous pneumothorax is a late complication of the disease. Herein we report a case of alveolar microlithiasis in a 42-year old male whose first presentation was the symptoms of pneumothorax. He was admitted with sudden onset dyspnea and right sided pleuritic chest pain. After treatment of pneumothorax with insertion of chest tube, pulmonary function revealed normal indices. The chest radiograph demonstrated diffuse confluence of dense micronodular infiltrate.

High-resolution CT scan revealed diffuse ground glass attenuation and calcifica-



tions along the interlobular septa and subpleural regions. A transbronchial lung biopsy confirmed the diagnosis of PAM.

P3506**Benign metastatic leiomyoma of uterus with lung damage**

Ivetta Dvorakovskaya, Dali Dzadzua, Irina Platonova, Laboratory of Pathomorphology, Research Institute of Pulmonology, St. Petersburg, Russian Federation

Benign metastasizing leiomyoma of uterus is difficult to diagnosis extrauterine metastatic leiomyomatous lung lesion without histologic signs of malignancy.

Aim: To present a rare case of uterus leiomyoma metastasizing into the lungs that shows difficulties in diagnosis and choice of adequate treatment.

Materials and methods: The woman, 46 year, was subjected to hysterectomy with uterine appendages in 2004. Through 5 years multiple masses were observed on the thorax x-ray that interpreted as lung metastases. With videothoracoscopy marginal resection of S4 on the right was performed. Histological sections were stained with hematoxylin-eosin and van Gieson. Immunohistochemical study was performed.

Results: Histological study revealed leiomyoma consisting of many small nodules of leiomyomatosis proliferation with multicellular foci, tendency to myxomatosis, and nuclear changes (polymorphism, blurred nuclear membranes, variety of chromatin content). Mitoses were absent. The histological structure of nodes removed from lungs and uterus was identical. High expression of antibodies to smooth muscle actin (SMA), expression of receptors Esr and Prs in uterine and pulmonary nodes indicated histological identity of the process that allows us to estimate changes in the lungs as metastasis. The patient was recommended hormone therapy (antiestrogen drugs in combination with tamoxifen).

Conclusion: Immunohistochemical study is necessary to determine the exact diagnosis of benign metastasizing leiomyoma of uterus with lung damage.

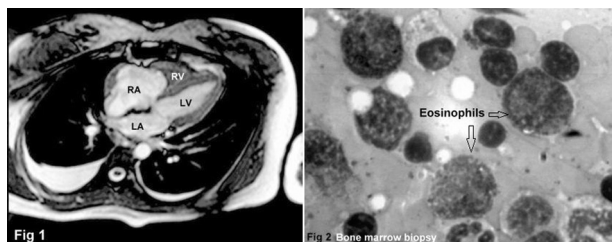
P3507**Hyper eosinophilic syndrome in a young female – A rare disorder**

Yannawar Anand¹, Ram Chopra¹, Nita Munshi², Nani Wadekar³, Vaibhav Pandharkar, Chintan Patel, Sharad Patil¹, Manisha Bhosle¹, ¹Chest Medicine, Ruby Hall Clinic, Pune, Maharashtra, India; ²Pathology, Ruby Hall Clinic, Pune, Maharashtra, India; ³Radiology, Ruby Hall Clinic, Pune, Maharashtra, India

Hypereosinophilic syndrome (HES) is characterized by blood eosinophilic count 1500 or more for 6 months & eosinophilic infiltrations of multiple organs like heart, bonemarrow, skin, CNS. We report a unique case, rare in a female

Case report: 20 years old female had progressive dyspnoea since 1 year. One report with her showed peripheral eosinophilia of 27% Respiratory system revealed signs of left pleural effusion with fluid eosinophilia Evaluated for this presentation Absolute eosinophilic count was 2750 cells. PFT, ECG were normal. CT thorax revealed RA thrombus & RV infiltrations and cardiac MRI showed typical endomyocardial fibrosis in RV.

Echo showed RA thrombus & apical fibrosis in RV. HES was considered strong possibility. Bone marrow showed 37% eosinophils, no blast cells.



Other causes of eosinophilia were ruled out. Diagnosis of HES was confirmed. BCR-ABL & FIP11-PDGFR-A fusion gene were negative. Patient responded to oral steroids & azathioprine.

Discussion: Multiple organ involvement with eosinophils is an important diagnostic criterion of HES. Malignant transformation is known. Early diagnosis, identification of disease variant are required to prevent fatal outcome.

TUESDAY, SEPTEMBER 4TH 2012

P3508**Bronchial artery aneurysm with hemothorax – A case report**

Catia Araujo, Natalia Andre, Maria Joao Tavares, Antonio Domingos.
Pneumologia C, Centro Hospitalar de Torres Vedras, Torres Vedras, Portugal

Bronchial artery aneurysm (BAA) is a rare condition (fewer than 40 cases have been reported in the literature), observed in less than 1% of all cases of selective bronchial arteriography. BAA is often diagnosed when it ruptures, with the most common symptoms being acute dyspnea, severe chest/back pain, hemoptysis/hematemesis (rupture into the lung/esophagus), symptoms of shock and sometimes hemothorax.

The authors report a case of a 65-year-old male patient, suffering of hypertension and permanent atrial fibrillation (treated with warfarin 5mg/day), with no previous history of pulmonary pathology, who presented with fever, wheezing, cough and hemopurulent sputum 4 days before. The chest radiography showed middle lobe (ML) and lower right lobe (LRL) consolidation leading to the diagnosis of community acquired pneumonia.

About 48 hours after the admission the patient had an episode of massive hemoptysis, severe dyspnea, wheezing and signs of shock, with a chest radiography showing a total right lung opacification. The bronchoscopy showed multiple clots in the ML and LRL, the right thoracentesis drained 2000cc of hematic fluid and the thoracic angio-computerized tomography documented an aneurysm of the right bronchial artery (8mm) and a contained rupture of a huge aneurysm of the intra-pulmonary branch of that artery (2.4cm) with intra-pulmonary hematoma and right hemothorax. Accordingly to the situation the patient was immediately submitted to surgery.

A massive hemothorax from a ruptured BAA is very rare. Though, this possibility must be considered in the absence of trauma history or other more common causes of hemothorax.

P3509**Respiratory findings of acromegalic patients**

Muhammed Emin Akkoyunlu¹, Fatmanur Karaköse¹, Mahmut Muzafer Ilhan², Levent Kart¹, Hatice Ozelik¹, Mehmet Bayram¹, Murat Sezer¹, Ertugrul Tazan². ¹*Chest Disease, Bezmialem Vakif University, Istanbul, Turkey;* ²*Department of Endocrinology, Bezmialem Vakif University, Istanbul, Turkey*

Acromegaly results from the long-term hypersecretion of growth hormone (GH) and elevation of levels of its peripheral mediator insulin-like growth factor-1. Acromegaly is known to significantly increase mortality rate. Cardiovascular diseases are the most common causes of death in acromegalic patients. Moreover acromegalic patients with cardiac hypertrophy have a decreased cardiopulmonary performance during exercise as compared to normal subjects. The aim of our study was to assess pulmonary functions and the cardiopulmonary response to exercise in acromegaly. Ten patients who were followed by endocrinology department outpatients clinic were enrolled to study. Disease activity was evaluated by growth hormone levels. All the patients were evaluated by the body measurements, pulmonary function tests, body plethysmography results. CPETs of the patients were performed. Ten patients (2 Male, 8 Female) were enrolled to study. Two of them were new diagnosed active acromegaly and 8 of them were under treatment. Mean age was 37.8±10.5. Two patients were evaluated for pulmonary function for the study before treatment while they were active disease duration. All the patient's FEV1/FVC ratio were greater than %70 (Mean: 80.7±8.8). All the TLC values of the patients were nearly upper limit of normal. When the CPETs were performed, 2 patients whose active disease test were ended because of the early arrhythmia and hypertension. None of the patients could reach maximal exercise capacity because of the muscular fatigue and all the patients VO2 max were reduced. Non invasive CPET results were compatible with poor effort.

P3510**A rare cause of hemoptysis: Munchausen syndrome**

Guzin Cinel¹, Suleyman Tolga Yavuz², Ugur Ozelik¹, Ebru Yalcin¹, Deniz Dogru¹, Nural Kiper¹. ¹*Department of Pediatrics, Pediatric Chest Diseases Unit, Hacettepe University, Ankara, Turkey;* ²*Department of Pediatrics, Pediatric Asthma and Allergy Unit, Hacettepe University, Ankara, Turkey*

Hemoptysis is a serious and sometimes life-threatening symptom that requires broad evaluation, including invasive procedures. Nevertheless in a group of patients, the cause of hemoptysis may not be elucidated despite extensive investigations. Factitious hemoptysis is a manifestation of Munchausen syndrome that describes a group of patients who intentionally produce or feign symptoms or disabilities; and it must be kept in mind in the differential diagnosis of unexplained hemoptysis. Here we report 5 patients diagnosed as Munchausen syndrome whose complaints were hemoptysis.

Patients: Five patients (1 male, 4 females), median age 15 (10-16,5), described hemoptysis occurring with cough or sputum. Symptom duration was 6 (1-24) months. They had not hematemesis or melena. Despite having hemoptysis everyday, or once in 2 or 3 days, hemoglobin values varied between 12.2-14.8 (median 13.2) g/dL. Chest x-rays, computed tomography of thorax, laryngoscopy, flexible bronchoscopy, hematological survey related with bleeding diathesis were normal in all patients. Accompanying psychiatric problems were determined in two patients with detailed history. In one patient, detailed physical examination revealed multiple crusted and bleeding bitched lesions on the buccal mucosa. One

patient brought us 3mL red fluid in a plastic cup as her bloody cough, but it was cherry juice. The other patient had problems with her school and family; and her hemoptysis was existing when she had bad notes at school.

Munchausen syndrome should be considered in the differential diagnosis of hemoptysis of unclear etiology, especially when accompanied by changing symptoms and in patients with a calm manner despite dramatic complaints.

P3511**Eosinophilic pneumonia – Our own experience with diagnosis and treatment**

Jan Plutinsky, Ivan Marget, Stanislav Majernik, Daniel Magula, Frantisek Dvorak. *2nd Pneumology, Specialized Hospital of St. Zorardus, Nitra, Slovakia (Slovak Republic)*

Eosinophilic pneumonia (EP) is a rare disease. Some EP could be caused by concomitant diseases (CD), while in others the cause is unclear (idiopathic). EP is divided into acute (AEP) and chronic (CEP) forms.

The aim: The authors share their own experience with EP diagnosis and treatment. **Methods:** EP was diagnosed in 23 patients (pts). Clinical examination, X-ray, CT, laboratory examinations with a peripheral blood count of eosinophils (Eo), microbiology, mycobacteriology, fiberbronchoscopy or histology were used. The pts were divided into two groups: AEP and CEP. Statistical analysis was performed.

Results: 15 F, median age 66 yrs (36-87) and 8 M, median 67 yrs (55-85) had one of the two forms: AEP in 7 pts and CEP in 16 pts. The main clinical symptoms were dyspnea in 20 pts (95.2%), cough in 19 pts (90.5%), fever in 17 pts (80.9%), pain in 5 pts (23.8%). AEP or CEP was the result of CD in 3 pts (toxoplasmosis), in 1 pt. with sarcoidosis, rhinitis and in 2 with allergic bronchopulmonary aspergillosis. In 17 pts no CD was identified. A statistical significance was found between the age of pts with AEP (56±12.2) and CEP (67.8±12.9) (p<0.05). There were no statistically significant differences between the absolute Eo count in M (614.8±279.3) and F (709.3±811.3) and in the groups of AEP (860.7±976.7) and CEP (579.6±429.3). 11 pts have been continually treated with corticosteroids, 8 pts temporarily. The longest treated patient has been receiving corticosteroid for more than 13 yrs.

Conclusions: EP is a very interesting disease. Treatment with antibiotics is unsuccessful, but corticosteroid treatment leads to a dramatic improvement. Dividing EP into AEP and CEP is not so clear cut.

P3512**Drug excipients allergy**

Vipulkumar Shah, Allergy & Asthma, Allergy Clinic, Surat, Gujrat, India

Introduction: Because of its rarity, drug excipients allergy is often unsuspected by the doctors. Some times the active drug is blamed and discontinued unnecessarily when the real culprit is drug excipients.

This was suspected when patients reported exacerbation of urticaria with anti histaminic which are actually used for treating urticaria. After suspecting drug excipient as the cause of allergy.

Material: To know actual cause it was decided to do patch testing with common drug excipients. The common excipient selected were Yellow Tartrazine, Titanium Dioxide, Sunset Yellow, Brilliant Blue, Quinolone Yellow, Talcum, and Ponacaeu 4R.

Method: Patch testing material were prepared by using the basic material and suitable vehicle. 24 patients were selected for patch testing. Patch was put on patients back after 48 hrs reading were taken to confirm the skin sensitivities.

Result: Out of 24 patients 15 patients were positive for Yellow Tartrazine only, 3 patients showed positive to Titanium Dioxide. 2 patients were positive to Quinolone Yellow and Talcum. 2 were positive with Brilliant blue. 3 patients were positive Yellow Tartrazine, Sunsets Yellow and Talcum.

Conclusion: Drug excipients play an important role in causation is required to be standardised for patch testing. This type of patients should be treated in view of such investigations and preferably treated with drug without excipients.

P3513**Rosai-Dorfman disease: Presentation of a clinical case**

Rubén Garrido¹, Elisa Barrera², Elías Abbud¹, Karla Cabada. ¹*Department of Pneumology and Thoracic Surgery, Specialty Medical Center, Juarez City, CHIH, Mexico;* ²*Institute of Biomedical Sciences, Autonomous University, Juarez City, CHIH, Mexico*

Introduction: Rosai-Dorfman disease (RDD) is a rare histiocytic proliferative process, infrequent and idiopathic, benign with painless cervical or systemic lymphadenopathy, fever, leukocytosis, polyclonal hypergammaglobulinemia and accelerated erythrocyte sedimentation. In a low percentage, there are extranodal manifestations. The histological diagnosis consists of filling of sinusoids in ganglia with lymphocytes, plasmatic cells and histiocytes positive for S100, CD68, absence of CD1a and emperipolesis.

Case report: 49-year-old female patient, who was a heavy smoker is presented with severe dyspnea, three months evolution and chest oppression sensation, with edema of face, neck and arms. The erythro sedimentation was accelerated (90 mm/hr). Chest x-rays showed mediastinum widening and tumor in the pulmonary right hilum. The computerized tomography showed right hilum hyperdensity of 7 cm. The bronchoscopy with biopsy showed right bronchi infiltrate with tumoral

TUESDAY, SEPTEMBER 4TH 2012

aspect. The histopathological findings revealed histiocyte proliferation and others with eosinophilic cytoplasm with fibrosis and lymphoid cells. Emperipolesis and plasmatic cells were identified, with Russell bodies. With the diagnosis 120 mg a day of 'attack dose' was initiated, continuing with 40 mg/day during 6 months with favorable response reducing the size of the lesion.

Discussion: In this case, the manifestation of the disease was dyspnoea produced by the bronchial obstruction and compression of the superior vena cava due to the hilum infiltrative histiocytic mass, so our initial probable diagnosis was a bronchogenic cancer. This extranodal manifestation is infrequent, but it is to be taken into account as part of the pulmonary and mediastinal pathology.

P3514

Three tumors in a young woman with Cushing's syndrome

Lizza Hendriks¹, Remy Mostard¹, Eric van Haren¹, Linda van de Winkel², Patrick van Battum², Wendy Schreurs³. ¹Pulmonary Medicine, Atrium Medical Centre, Heerlen, Netherlands; ²Internal Medicine, Atrium Medical Centre, Heerlen, Netherlands; ³Nuclear Medicine, Atrium Medical Centre, Heerlen, Netherlands

Case: A 38-year old female with no relevant medical history was referred to internal medicine with clinical features comparable with Cushing's syndrome. Laboratory results suggested ectopic adrenocorticotrophic hormone (ACTH) production. The pituitary gland had a normal aspect on magnetic resonance imaging but computed tomography of the chest showed a smooth, oval mass in the posterior mediastinum and a nodule in the left upper lobe. Differential diagnosis consisted of carcinoid, small cell lung cancer or neurogenic tumor. Bronchoscopy showed no endobronchial lesions but both lesions could not be reached with fluoroscopic guidance. On ¹⁸F-deoxyglucose positron emission tomography (¹⁸FDG-PET) increased ¹⁸FDG uptake was seen in the mediastinal mass and in the right gluteal region, but not in the left upper lobe. Somatostatin scintigraphy showed uptake in the left upper lobe, but not in the other masses. Histological biopsy of the gluteal mass showed a desmoid type fibromatosis. Wedge resection with peroperative frozen section of the left upper lobe mass was performed and a typical carcinoid (T1a) was found. For treatment and staging resection of the left upper lobe with mediastinal lymph node sampling and resection of the mediastinal mass followed in the same session. The resections were complete but one lymph node (N2) was positive for carcinoid metastases. The mediastinal mass appeared to be a schwannoma.

Conclusion: Cushing's syndrome caused by ectopic ACTH in a T1aN2M0 typical carcinoid. Furthermore, the two PET-positive lesions appeared to be a schwannoma and a desmoid type fibromatosis. This case stresses the importance to link presenting condition with imaging features and to obtain histological confirmation.

P3515

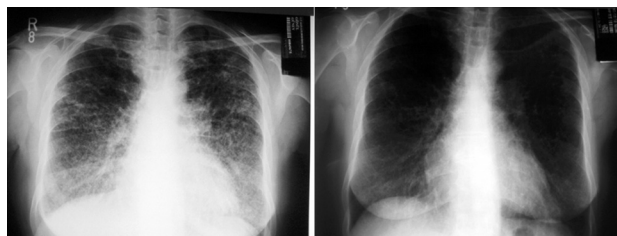
The treatment of a non-Langerhans-cell histiocytosis, Erdheim-Chester disease: A-case report

Dane Ediger¹, Asli Gorek Dilektasli¹, Ahmet Ursavas¹, Duygu Koprucuoglu¹, Saduman Balaban Adim², Gokhan Gokalp³, Ugur Yayla⁴, Ercument Ege¹.

¹Department of Pulmonary Diseases, Uludag University Faculty of Medicine, Bursa, Turkey; ²Department of Pathology, Uludag University Faculty of Medicine, Bursa, Turkey; ³Department of Radiology, Uludag University Faculty of Medicine, Bursa, Turkey; ⁴Department of Ophthalmology, Uludag University Faculty of Medicine, Bursa, Turkey

Erdheim-Chester disease (ECD) is a rare, non-Langerhans histiocytosis with multi-system involvement. Pulmonary involvement is uncommon. We present a 46-year-old woman who presented with chronic cough, dyspnea and yellowish plaques in the periorbital area. She was previously diagnosed as diabetes insipidus. Chest radiographs showed bilateral diffuse interstitial infiltrates.

Symmetric sclerotic bone lesions, dural nerve thickening were defined on imaging studies. Histopathologic examination of the skin lesions revealed infiltration of CD-68 positive foamy histiocytes. She was diagnosed as ECD with pulmonary, pituitary, skeletal, orbital, central nervous system and skin involvement. Following first-line treatment with corticosteroids her symptoms worsened. Treatment with interferon-alpha was started. She had an marked improvement in symptoms, radiologic findings and skin involvement under this treatment.



Conclusion: ECD should be considered in the differential diagnosis of interstitial lung diseases. As there is no standard treatment for this disorder, interferon-alpha can be effective in the treatment of Erdheim-Chester disease.

P3516

High grade laryngeal stenosis caused by an extramedullary metastasis of plasmacytoma

Joern Brune. *Pneumology, Lung Centre Hemer, Hemer, Germany*

We report the case of a 69-year-old woman who presented to our department with rapidly progressive hoarseness, stridor and dyspnea for week. The patient had a history of multiple myeloma for 3 years and resected breast cancer for 1 year.

On fiberoptic bronchoscopy a high grade (>80%) right sided laryngeal submucosal mass, affecting both supra- and infraglottic parts of the larynx, was found the reason for her symptoms. We performed a surgical tracheostomy considering it the only viable option of airway management.

An MRI scan of the neck revealed a right sided cervical mass of 7x3cm, as well as an enlarged lymph node in the right supraclavicular region (1R). Biopsy was performed both of the lymphnode (core needle biopsy) and the laryngeal mass (EBUS-TBNA). The histological and immunohistochemical findings were consistent with metastasis of the known multiple myeloma.

Extramedullary metastasis of multiple myeloma is a rare complication of the disease occurring. Laryngeal stenosis due plasmacytoma has been described in pediatric patients, but never in adult patients before.

P3517

Urinary incontinence in adult bronchiectasis patients: Common but treatable

Jonathan Miller¹, Hilary Tedd¹, Jackie Rees², Anthony DeSoyza¹.

¹Bronchiectasis Service, Newcastle NHS Foundation Trust, Newcastle, United Kingdom; ²Continence Service, Newcastle NHS Foundation Trust, Newcastle, United Kingdom

Background: Patients suffering from urinary incontinence (UI) often avoid seeking medical attention, due to embarrassment, consequently remaining untreated. UI negatively impacts of quality of life, psychological health and relationships. In patients with non-CF bronchiectasis, it may also reduce adherence to chest physiotherapy. UK Physiotherapy Guidelines (2009) suggests screening for UI in patients with non-CF bronchiectasis.

Methods: Screening at new specialist bronchiectasis clinic for symptoms of UI, with referral onto Continence specialist services.

Results: Of the initial 116 patient referred to the bronchiectasis service, 76 were female. 55% of female patients had UI (UI-Br). 87.5% of UI-Br patients reported symptoms for over 5 years, with 40% of patients describing symptoms of over 10 years duration. 37% reported UI as having a terrible impact on quality of life. Patients were assessed by a Continence Nurse Consultant. A personalised UI management plan was formulated, including education on pelvic floor strengthening, urge suppression and voiding techniques. Other techniques used included bladder retraining (40%) and toilet rescheduling (40%). Over 60% of patients have been discharged by the Continence Service, following symptom improvement.

Conclusions: In asking about UI, in line with UK guidelines, we identified symptomatic patients and referred them onto an appropriate specialist service. We identified a high prevalence of UI in female patients with non-CF bronchiectasis. Patients suffered symptoms for a prolonged period with a significant impact on quality of life. Through appropriate specialist input, these distressing symptoms have been improved in the majority of our patient group.

P3518

Hypogammaglobulinemia, reduced B cell count and recurrent sinopulmonary infections: Good's syndrome

Murat Türk, Nurdan Köktürk, Numan Ekim. *Pulmonary Medicine, Gazi University School of Medicine, Ankara, Turkey*

A 62-year-old woman with a history of thymectomy for capsule-invasion-free, type AB thymoma, presented with cough, dyspnea and fever. She had frequent respiratory tract infections for 9 months. Her initial physical examination was normal. Abnormal laboratory studies on admission were anemia, leukocytosis, proteinuria, elevated erythrocyte sedimentation rate, C-reactive protein and procalcitonin levels. On chest tomography, multiple mediastinal and hilar lymph nodes, bilateral traction bronchiectasis and millimetric nodules were seen. E.Coli was isolated in bronchoalveolar lavage. TBNA and TBB were negative for probable thymoma metastasis.

For the probability of Good's Syndrome, serum immunoglobulin levels were measured and found normal for IgA, low for IgG and high for IgM. Flow cytometry demonstrated 8% of CD19+, 12% of CD56+, 14% of CD57+, 30% of CD4+, 37% of CD8+ cells in total lymphocytes; consistent with B-cell immunodeficiency. With the diagnosis of Good's Syndrome, intravenous immunoglobulin treatment was started. During her follow-up, she was hospitalized twice for respiratory tract infections. Now she is on ciprofloxacin prophylaxis.

Good's syndrome is a rare cause of combined B or T cell deficiency. Its main characteristics are hypogammaglobulinemia, reduced or absent B cells, reduced serum levels of IgG, IgA and IgM, recurrent sinopulmonary infections caused by encapsulated microorganisms. Management of syndrome includes surgical resection of thymoma, treatment of infections and immunoglobulin replacement therapy.

Good Syndrome is a rare but treatable condition and it should be considered in patients with the diagnosis of thymoma and frequent respiratory tract infections.