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84. Rare clinical cases

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Williams-Campbell syndrome: is an infantile rare disease with poor prognosis?

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Williams-Campbell syndrome, described by Williams and Campbell in 1960, is a rare disorder characterized by abnormal ballooning and collapse of the affected bronchi. As early descriptions were made in children, this disease has been believed to occur from congenital deficiency of bronchial cartilages and have poor prognosis.

We have experienced 15 adult cases of Williams-Campbell syndrome (male:female=8:7) from 1988. All of them were diagnosed by either bronchography or chest CT. In this presentation, we reviewed focusing on the age distribution, respiratory infection, pulmonary function, and vital prognosis of these cases.

As a result, average age of these patients was 63.6 (from 48 to 76). Past history of pneumonia was seen in 10 cases, positive sputum culture in 13 cases (especially *H. influenzae* and *P.aeruginosa*), and restrictive impairment of pulmonary functions in 12 cases. Two patients died of asthmatic attack and respiratory failure. 6 of 8 traceable cases are alive after the follow-up of 4 to 14 years. Surgeries have been successfully performed for 1 case with uncontrollable hemoptysis and 2 cases with malignancies from the affected bronchi (lung cancer and malignant lymphoma).

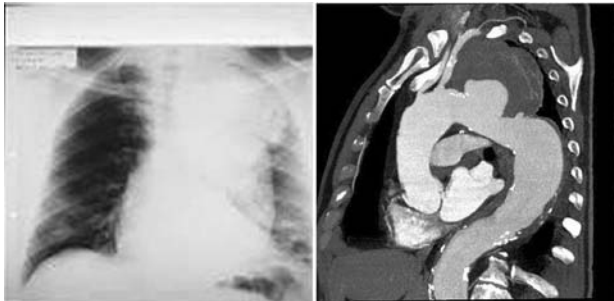
In conclusion, we assumed that Williams-Campbell syndrome might be seen among the elderly, and the prognosis of this disease might be better than previously reported. Malignancies were seen in two patients, though the correlation between the malignancies and Williams-Campbell syndrome is unclear.

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Unusual cause of haemoptysis – case report

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Haemoptysis, especially when massive, is a life-threatening symptom, an early assessment of the likely underlying cause needs to be made and the specific treatment. A 66 years old patient, with cardiovascular risk factors, diagnosed 3 years ago as aortic aneurysm with stationary development. He accused dyspnoea, fatigue, atypical chest pain and haemoptysical sputum in small quantity. The ECG and the laboratory investigations do not reveal important changes, but the X-ray chest shows, beside the descending aorta dilatation, an opacity without defined boundary-line, relatively homogeneous, in the upper left lobe.



He is under suspicion of lung cancer, but the CT appearances reveals a giant aneurysm, in the left upper lobe.

After an arduous anamnesis, the patient mentions that 3 months ago in the middle of a road transport breakdown he was suffering a violent trauma that didn't affect him then. Catheterization reveals ascending aorta dilatation, without lesions of the big vessels from the aortic arch. The aortic arch reveals important aneurismal dilatations and return at its normal shape above diaphragm. He leaves hospital with an appointment for a surgical procedure.

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PAS-negative pulmonary alveolar proteinosis

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A 30 yr old male patient presented with persistent cough for 1 year and drop in effort capacity for few months. Before presenting to our center, restrictive lung

functions and diffuse ground glass appearance on HRCT were found with non-specific BAL findings. He was started on methyl-prednisolone for 6 weeks with no radiological improvement and clinical progression. Upon presentation, he had good general condition, stable vital signs and bilateral crepitations on auscultation with otherwise normal physical examination and chemistry. Pulmonary function tests showed a FVC and DLCO of 64% and 37% of predicted respectively. BAL material was milky and microscopy revealed eosinophilic amorphous material not stained with PAS and Grocott, which also filled the alveolar spaces on transbronchial lung biopsies. All microbiological studies were negative, and PAS negative PAP diagnosis was established, all medications were stopped and total lung lavage was performed for both lungs within 2 weeks period under general anesthesia through a double lumen endotracheal tube using normal saline at body temperature with a total of 20 liters for the right lung and 16 liters for the left lung. Ten days after the procedure the patient was evaluated to reveal significant clinical improvement, with increase in FVC and DLCO to 73% and 47% of predicted respectively. A 19% increase in 6 minutes walking distance with no accompanying hypoxemia upon effort. Following HRCTs showed decrease in the ground glass appearance with stability upon succeeding studies. He is being followed up for the last 9 months with no need for another lavage. This is a case of PAP with typical clinical, radiological and functional findings but with PAS negative eosinophilic alveolar deposits.

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The respiratory manifestations of Behcet disease: a study about 26 cases colligated in 12 years (1995-2007)

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Background: In 1937, Heluci Behcet, a Turkish dermatologist described a disease associating mouth and genital ulcers, together with uveitis. Nevertheless rare, the mediastine- pulmonary localizations made by vascular thrombosis and pulmonary vasculitis are serious.

Objectives: The aim of this study is to point out the rarity and the gravity of the respiratory manifestations of the Behcet disease, and suggest a therapy framework.

Methods: Analysis of 26 cases of Behcet disease with mediastine-pulmonary manifestations colligated at our department between 1995 and 2007.

Results: The mouth ulcer has been observed in all patients whilst genital ulcer in 15, ocular localizations in 9. In 9 patients, vascular manifestations has been noticed 10 times (peripheral phlebitis: 7 cases and venous thrombosis: 3 cases). An articular localization has been observed in 8 patients (Arthralgia and arthritis). The diagnosis is based essentially on medical scanning. The treatment depends on the existence of superior vena cava thrombosis, or a pulmonary vasculitis. As, there is no codified therapeutic protocol, the treatment is prescribed almost obeying to the case by case rule. It includes: Corticoids and/or anti-coagulants and/or immunosuppressants. The duration of the therapy is modulated according to the results of the prescribed treatment.

Discussion and comments: The mediastine-pulmonary localizations of the Behcet disease represent a preoccupying entity by its clinical seriousness and its therapeutical difficulties. On the basis of our experience, we suggest a taking care plan of this serious and capricious disease which treatment and its results are generally disappointing.

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Lymphangioliomyomatosis: characteristics of 96 patients in China

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Background: Pulmonary lymphangioliomyomatosis (LAM) is a rare disease that is associated with infiltration of atypical smooth muscle-like cells. Sporadic LAM may have pulmonary involvement and extrapulmonary manifestations including renal angiomyolipomas, axial lymphadenopathy, or abdominal lymphangiomyomas. Increased recognition of LAM and widespread availability of imaging techniques ensure that more LAM patients are diagnosed in China.

Methods: Eleven newly reported patients with sporadic LAM integrated with 85 sporadic LAM published in the Chinese literature over a 13-yr period, from 1993 to 2007 were reviewed.

Results: All 96 patients were women aged 13 to 56 yr (mean±SD, 37±8.4 yr). The average age at onset of symptoms was 34.3±7.6 yr. Pulmonary manifestations, most commonly exertional dyspnea were the primary events leading to the diagnosis in 83.3% of cases. The most common pulmonary function abnormality is airflow limitation observed in 60% of the patients, whereas only 6.9% had normal spirometric results. 74% of the LAM patients have a reduced diffusing capacity. Renal angiomyolipomas were found in 30% of the patients. 40% of the patients were treated with a progesterone derivative with little evidence of efficacy. The disease with deterioration was observed in 23% of the patients over a 3-months' to 5-yrs' follow-up.

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Conclusions: The age range and the degree of pulmonary function afflicted with LAM can be quite variable. The pulmonary dysfunction in our group of LAM patients likely reflects the clinically significant respiratory symptoms when diagnosed.

P561**Successful treatment of Churg-Strauss syndrome with cyclosporin after oral steroids discontinuation due to side effects**

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Churg-Strauss syndrome is characterized by history of asthma, peripheral blood eosinophilia and tissue eosinophilia with predilection to lungs. We would like to present the case of 35 year-old patient with severe asthma. On admission to the hospital the patient was in severe state, physical examination suggested bilateral pneumonia, confirmed by chest X-ray, blood gas analysis showed respiratory failure. The patient was successfully treated with three antibiotics. On subsequent hospitalization one month later, he presented with fever, cough, dyspnea, myalgia and symptoms of sinusitis. Chest X-ray revealed pulmonary infiltrates, peripheral blood smear showed eosinophilia 50%. Further investigations revealed negative test for both antineutrophil cytoplasmic antibodies. The therapy with oral steroids was started with resolution of symptoms. After 10 months of oral steroids use, the patient again developed dyspnea, fever, obturation, hypoxemia, infiltrates seen on chest X-ray. The diagnosis of Churg Strauss Syndrome was established on clinical basis and cyclosporin was added to the corticosteroids. Clinical improvement in the patient's general health and normalization of laboratory tests were achieved again. The patient was under surveillance for 2 years and over this time symptoms did not recur. Cyclosporin was discontinued after 12 months of use. The patients decided to withdraw oral steroids because of the whole range of adverse symptoms. The new regimen with cyclosporin 2 x 320 mg was introduced. Today the patient is in good health with no symptoms, the laboratory findings show eosinophilia of 6%, which is the lowest in his disease course.

P562**A 28-year-old man with dyspnea and generalized edema**

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A 28-year-old man was referred to the hospital in February 2005 because of elevated liver enzymes (AST, 192 IU/ml; ALT, 202 IU/ml) without symptoms. The serologic tests for Hepatitis B virus (HBV) markers, anti-HAV antibody, anti-HCV antibody, antinuclear antibody, anti smooth muscle antibody, and anti mitochondria antibody were normal. Sonographic finding of the liver was also non-specific. After the first visit, the patient failed to follow-up. One year later, he presented with generalized edema especially face and legs for 3 months. The urine was positive for protein (+++); the sediment contained many red cells. AST and ALT were 127 and 91 IU/ml, respectively. Kidney biopsy showed membranous glomerulonephritis, with focal endocapillary proliferation and global sclerosis (5.3%) and segmental sclerosis or hyalinosis (47.4%). Liver biopsy showed chronic hepatitis with mild lobular activity, mild porto-periportal activity and periportal fibrosis. The generalized edema improved with diuretics. In October 2007, he was admitted because of generalized edema and dyspnea. Pulmonary function testing showed moderate restrictive pattern. Chest radiology showed multifocal nodular consolidation with air-bronchogram and patchy ground glass opacities. Open lung biopsy from the right middle lobe showed pleural and perivascular histiocytic infiltration. The immunohistochemical stains study suggested the histiocytes are not originated from the Langerhans cell and S-100 protein and CD1a negative. He was diagnosed as Erdheim-Chester disease. He was treated with steroid combined with cyclosporin. Unfortunately, his general condition deteriorated. He required mechanical ventilation but died of respiratory failure on day 38 after admission.

P563**Invasive aspergillus tracheobronchitis (AT) in non-neutropenic, immunocompromised patients**

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Introduction: Ulcerative and pseudomembranous tracheobronchitis are rare, invasive forms of tracheobronchial aspergillosis.

Objectives: To evaluate the incidence, clinical aspects and outcome of invasive AT in non-neutropenic, immunocompromised patients.

Material: 49 consecutive, non-neutropenic, immunocompromised patients (mean age 57; range 29-79 yrs) with clinical and/or radiological symptoms of pulmonary infection.

Methods: All patients underwent clinical assessment, thorax CT scan and fiberoptic bronchoscopy with bronchoalveolar lavage (BAL). Cultures, Aspergillus galactomannan (A-GM) detection and PCR methods were used to search for Aspergillus spp. in blood and BALF.

Results: In 3 patients (3/49; 6%) endobronchial picture consistent with ulcerative and/or pseudomembranous tracheobronchitis was found. This was confirmed by endobronchial biopsies, which showed fungal invasion of the bronchial wall. Cough, hoarseness and fever were dominating clinical symptoms in these patients. A-GM in BALF was positive in 2 patients. In these patients AT was associated with invasive pulmonary or disseminated aspergillosis. They died despite of intensive antifungal treatment. In one patient with local tracheobronchial involvement both serum and BALF A-GM studies gave negative results.

Conclusions: Invasive AT might be the cause of pulmonary syndromes in non-neutropenic, immunocompromised patients. When associated with pulmonary and/or disseminated diseases, the prognosis is poor. BALF and serum A-GM detection has limited diagnostic value in patients with AT.

P564**Tracheobronchial straight metallic pin aspiration (SMP) in young Jordanian females**

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Background: SMP (used for securing facial and headscarfs) aspiration has been reported as a new form of FB aspiration in Islamic countries.

Objectives: To define the clinical spectrum of tracheobronchial SMP aspiration in adults and evaluate the efficacy of bronchoscopy at the largest two hospitals in Jordan.

Methods: Retrospective analysis of 73 consecutive clinical series of adult patients from two referral-based medical centers (40 patients at King Hussein Medical Center (KHMC) and 33 patients at Al-Basheer Hospital) over seven years period (July 2000- July 2007) were analyzed. The Flexible Fiberoptic Bronchoscopy (FFB) was used for FB identification and removal in all patients who presented to KHMC, video assisted rigid bronchoscope under general anesthesia was used at Al-Basheer Hospital.

Results: All patients were females below 30 years of age. 69 patients (94.5%) presented within the first 24 hours after aspiration. Medical history was suggestive of FB aspiration in all patients. Chest X-Rays demonstrated the radio opaque pins in all patients. At bronchoscopy the intermediate bronchus was the most common site of FB lodgment (64%). In all KHMC patients, the aspirated pins were removed successfully from the first attempt. The procedure had to be repeated in 4 cases of Al-Basheer hospital patients. A thoracotomy was performed in another 2 patients after unsuccessful interventions. 90% were discharged on the same day of the procedure. All patients has uneventful recovery.

Conclusion: Tracheobronchial SMP are relatively frequent FBs in Jordanian young females and can occur in a very specific clinical setting. Removal of these FBs can usually be accomplished successfully by FFB.

P565**A rare case of lithoptysis in a patient with Kartagener's syndrome**

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Introduction: Lithoptysis is characterized by the expectoration of bronchial calculi (broncholiths) that are defined as calcified material in the lumen of the tracheobronchial tree. Case reports showing an association between lithoptysis and Primary Ciliary Dyskinesias (PCD) are rare and was first described by Kennedy et al¹ in 2006. We would like to add one more case to this rare series.

Case report: A 48 year old male ex-smoker, with severe bronchiectasis and obstructive airways disease (FEV₁ 0.67 l) secondary to Kartagener's Syndrome, was under regular review in the Respiratory clinic. In Oct 2006 he reported several episodes of expectorating sputum mixed with gritty material. On one occasion he brought two large pale coloured broncholiths that he had coughed up, to clinic. Biochemical analysis confirmed these as calcium oxalate stones.

Discussion: Kartagener's Syndrome is characterized by the clinical triad of chronic sinusitis, bronchiectasis, and situs inversus. It is part of the larger group of disorders referred to as PCD. Symptoms result from defective cilia motility. The precise nature of stone formation in the bronchial lumen is unknown.

The calcite stone formation is hypothesized to be due to a biomineralization response to chronic airway inflammation and retention of infected airway secretions in PCD patients. Another hypothesis is the erosion of calcified granulomas/lymph nodes into the bronchial lumen.

The most common causes of broncholiths are tuberculosis and histoplasmosis, but can also arise in other situations, such as aspiration of bone tissue, calcification *in situ* of an aspirated foreign body, erosion and extrusion of calcified bronchial cartilage, and silicosis

Reference:

1. Respir Med. 2006 Jun 4; 167:571-59.

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P566**Primary cardiac angiosarcoma and diffuse pulmonary hemorrhage: case report**Melih Buyuksirin, Gulru Polat, Adnan Usalan, Gultekin Tibet. *Dr Suat Seren Chest Diseases and Surgery Research and Training Hospital, Izmir, Turkey*

Primary cardiac angiosarcoma is rare malign tumour with very poor prognosis. Symptoms are nonspecific. One of rare clinical presentations is diffuse pulmonary hemorrhage. 61 year old man applied with cough and hemoptysis. The chest Xray showed bilateral nodular infiltrates and increase in cardiothoracic index. Mass in 7x5 cm dimensions in the right atrium (myxoma?, thrombus?) and ground glass appearance on the middle and lower zones observed on the thoracic computerized tomography (CT). Left ventricle was dilated and hypertrophic on echocardiography (ECHO). The case was operated with the prediagnosis of myxoma. Cardiac angiosarcoma identified in the operation. We presented the case because of the difficulties in the diagnosis, being diagnosed by pulmonary symptoms and died of respiratory failure in a short time.

P567**A case of toxocarasis diagnosed as bronchiolitis obliterans with organizing pneumonia**Rosanna Qualizza, Cristoforo Incorvaia. *Allergy, Istituti Clinici di Perfezionamento, Milan, Italy; Allergy/Pulmonary Rehabilitation, Istituti Clinici di Perfezionamento, Milan, Italy*

Background: *Toxocara canis* is a nematode affecting dogs and cats, humans are hosts in whom the larvae do not develop to adult worms but may migrate in various tissues and organs, associating to a number of clinical symptoms concerning the skin, the eye, and the lung, which may lead to wrong diagnosis.

Methods: Here we report the case of a patient diagnosed with bronchiolitis obliterans with organizing pneumonia (BOOP). He referred to us when 57 years-old because of persisting dermatitis, but had had repeated hospitalisations for cough and dyspnea with radiographic infiltrates. In 2000 a chest TC was indicative of BOOP and a further hospitalisation with the same diagnosis occurred in 2002, with concomitant spirometric findings of asthma-like obstructive respiratory deficit.

Results: The patient referred to us for allergy evaluation in February 2004. Skin tests gave negative results, and a complete blood examination with parasitologic testing was suggested. At the control visit all results were negative but Western blotting (WB) for *Toxocara canis*, with a negative ELISA. Treatment with mebendazole repeated for three courses at monthly interval resulted in remission of skin symptoms and significant improvement of lung symptoms and spirometric values. In 2005 a relapse of dermatitis, but not of lung symptoms, along with a persistent positivity of WB required further three courses of mebendazole. At the latest control in October 2007 the patient was asymptomatic and WB was negative.

Conclusion: The present case suggests to consider the possible responsibility of *Toxocara canis* in patients with complex lung symptoms and signs including asthma (as already described) but also BOOP, thus far unreported.

P568**Pulmonary hypertension due to ligation of pulmonary arteriovenous malformations**Zeynep Pinar Onen, Gozde Koycu, Gulseren Karabiyikoglu. *Department of Chest Diseases, Ankara University School of Medicine, Ankara, Turkey*

Pulmonary arteriovenous malformations (PAVMs) are abnormal vascular structures that direct capillary-free communication between the pulmonary and systemic circulations. PAVM has been described in all ages but most patients present in the second decade of life. In these chronically adapted patients, there is a reduced total pulmonary vascular resistance, normal mean pulmonary arterial pressure (PAP) and increased cardiac output.

24 year-old man who had complained of dyspnea on exertion for a few last years referred to physician. Chest x-ray revealed a nodular lesion on the right and computed chest tomography demonstrated it as sub pleural vascular lesion compatible with arteriovenous malformation. Before the operation systolic PAP with echocardiography was 40mmHg and the arteriovenous malformation was ligated. 4 months after the operation; he admitted to our clinic with progressively increased dyspnea on exertion and hemoptysis. On his physical examination common systolic murmur heard on chest wall with a systolic murmur of heart. Echocardiography revealed systolic PAP of 150mmHg and pulmonary artery angiography analyses detected compatible appear with common arteriovenous malformation.

Treatment options for PAVMs include embolotherapy and operation. However, surgery was never the ideal solution for the multiple PAVMs because these are low resistance shunt areas and ligation of them may cause or aggravate the pulmonary hypertension. Lung transplantation may be a better choice for the patients with multiple PAVM whom had negative response to embolotherapy.

P569**A case of bronchial anthracofibrosis**Prakash Gupta, Milan Bhattacharya, Vijayadwaja Desai, Rahul Mukherjee. *Respiratory Medicine, Milton Keynes Hospital, Milton Keynes, United Kingdom*

Background: The bronchoscopic appearance of narrowing or obliteration of the

bronchial lumen associated with anthracotic pigmentation without a relevant history of smoking or coal dust exposure has been defined as anthracofibrosis.[1] Identified about 10 years ago, it occurs mainly in elderly Asian women and is associated with previous tuberculosis (TB).

Case Report: An 87 year old Indian housewife presented with 1 week of dyspnoea and cough with purulent sputum. Her past medical history included pulmonary TB treated in India nearly 50 years ago, breast cancer for which she was on Tamoxifen, hypertension, type 2 diabetes requiring insulin and depression. She never smoked, drank alcohol or had coal dust exposure. She was severely anxious and tachypnoeic. She had bilateral crepitations but was not clubbed. A high resolution computed tomography (CT) of the thorax showed consolidative changes particularly in the right hilum with narrowing of the adjacent segmental bronchi. Bronchoscopy showed narrowing of the bronchial lumen in the right middle, upper and left upper lobes with anthracotic pigmentation. Bronchial biopsy confirmed sub-epithelial chronic inflammation with pigmentation. Bronchial lavage grew *Aspergillus fumigatus* but was negative for TB. She recovered after 4 weeks of oral Itraconazole 200mg per day, suggesting that the acute illness was possibly due to aspergillus bronchitis supervening on bronchial anthracofibrosis.

Conclusion: This case demonstrates that bronchial anthracofibrosis is a diagnosis to be considered in this patient group with previous TB and characteristic CT and bronchoscopic appearances.

1. Chung MP, Lee KS, Han J, et al. Bronchial stenosis due to anthracofibrosis. *Chest* 1998;113:344-350.

P570**Paraneoplastic syndrome in 70-year old male? A rare presentation of *Streptococcus intermedius* (SI) infection**Ali Raufi¹, Muhammad Ali², Alia Waheed³, James Rowley², Renee Dwaihy¹.

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70 yo male was admitted with intermittent confusion, loss of balance, eyelid droop, positional dizziness, & a 25 lb weight loss over 3 months. He denied hemoptysis, fever, chest pain, or loss of consciousness. He was hospitalized in Missouri for progressive neurologic symptoms but had negative work-up. CT thorax there revealed an incidental 6 cm right (Rt) lung mass.



Medical history: Hypertension, hyperlipidemia, atrial fibrillation. He smoked 5 cigar/day for 15 yrs but quit 10 yr ago. He is a retired engineer without occupational exposures. Medications: fluvastatin, diltiazem, lisinopril, cyclobenzaprine, trazodone & fondaparinux.

Neurologic work-up (Head CT, MRI, LP, Physical exam) was unremarkable. CT guided biopsies of the mass revealed inflammatory cells. PET scan revealed an FDG-avid mass & a hilar lymph node. High clinical suspicion of malignancy led to Rt upper lobectomy, which showed no malignant cells but tissue cultures grew SI. The patient responded to amoxicillin/clavulanic acid, & was discharged home. SI, a member of the *S. milleri* group, very rarely causes infections of the lower respiratory tract without clinical respiratory symptoms (R. Whitley et al; JCM 1992). Our case is unique because the clinical presentation raised suspicion of lung malignancy with a paraneoplastic syndrome. Elderly patients with such chronic infections may present with a wide spectrum of clinical findings & require careful evaluation.

P571**Rhabdomyoma as a tumor of the posterior mediastinum**Joanna Kuschił-Dziurda, Lucyna Mastalerz, Piotr Grzanka, Ewa Nizankowska-Mogilnicka. *Department of Pulmonary Diseases, Jagiellonian University, School of Medicine, Cracow, Poland*

Rhabdomyoma is a benign neoplasm of skeletal muscle occurring mainly in the head and neck region. Rhabdomyoma is extremely rare cause of a tumor in the mediastinum. We present a case of an adult rhabdomyoma, which is located multifocally in the head, neck and mediastinum. There have been only four previous reports of a rhabdomyoma in the mediastinum. A 80-year old, white man was admitted to our Department of Pulmonary Diseases with dyspnea, haemoptysis, cough, profuse sweating, fever and weakness. Physical examination revealed tumor in left submandibular area of the neck. Chest radiography showed no changes. Neck computed tomography (CT) showed the mass causing prominence of the left

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postero-lateral wall of the pharynx. This mass measured 52x24x58mm and enhanced with contrast medium (120 HU). Chest CT revealed tumor (20x19x55mm) which was in contact with the lateral wall of the esophagus and similarly enhanced with contrast medium. Histologically, the tumor was composed of the adult rhabdomyoma cells. The multifocal localization and benign character of the tumor were contraindication to surgery. To our knowledge, this is the fifth case of an adult rhabdomyoma occurring in the mediastinum. During the following six years the tumors did not progress (stable in CT). Currently the patient complains of productive cough, mild dyspnea, dysphagia and hoarseness.

P572**Brown tumor of the rib – a case with parathyroid adenoma**

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Brown tumor can be a rare skeletal manifestation of advanced hyperparathyroidism. Brown tumor arises as a direct result of the effect of parathyroid hormone on bone tissue in patients with hyperparathyroidism.

We reported a case of brown tumor of the rib with a preoperative incident finding in chest film of a 23-year-old man with chronic renal failure. Chemical blood analysis revealed increased serum calcium levels and the parathyroid hormone level. Patient was initially accepted as loculated pleural effusion. But ultrasonography revealed no pathology. In computerized thorax tomography, there was 3x5 cm oval shaped mass lesion in lateral part of 3. and 5. rib. The patient underwent surgery with removal of a parathyroid mass.

Brown tumors are rare complication of hyperparathyroidism and should be considered in the evaluation and differential diagnosis of patients with atypical presentation.

P573**Mediastinal lymphadenopathies: a rare location of amyloidosis**

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Introduction: Amyloidosis is exceptionally revealed by mediastinal lymph node enlargement caused by amyloid deposits.

Case report: This study examined the case of a 64-year-old male, smoker of 45 packets per year, without any past medical history, presented with complaint of dyspnea and dry cough during the last two months. Upon physical examination he had superior vena cava syndrome.

The bronchial fiberoptic showed narrow left and right upper bronchi. Chest CT-scan revealed several mediastinal lymphadenopathies. Abdominal CT-scan was normal.

The histological study with Congo red stain of a mediastinal lymph node biopsy determined the diagnosis of amyloidosis. Abdominal ultrasonography was normal. Biological examination determined a normal renal function with creatinemia of 65 µmol/L, without any nephrotic syndrome. The final diagnosis was idiopathic amyloidosis. One year after the diagnosis the evolution has been marked by clinical stability without any systemic dissemination notably cardiac, hepatic or renal locations.

Conclusion: Although it is a rare occurrence, amyloidosis should be taken into consideration in the differential diagnosis of mediastinal lymphadenopathies. In cases of uncertain diagnosis Congo red stain among others, and an immunohistochemical study should be performed.

P574**An unusual case of catamenial pneumothorax (CP)**

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CP is a rare cause of spontaneous pneumothorax (SP). It is the most common form of thoracic endometriosis syndrome, which also includes catamenial haemothorax, catamenial haemoptysis and endometriosis lung nodules. It is defined as a recurrent syndrome of pneumothorax that occurs between 48 and 72 hours after menstruation. It accounts for 2.8 to 5.6% of all SP episodes in women. Most patients have a history of pelvic endometriosis and often have recurrent pneumothoraces before it is recognised. We report a patient who failed thoracic surgical intervention and was subsequently successfully treated by hormonal treatment, followed by Total Abdominal Hysterectomy (TAH) and Bilateral Salpingo Oophorectomy (BSO).

Case report: A 35 year old woman presented with right sided chest pain in 2001. Chest x-ray confirmed a right sided SP which was successfully managed by aspiration. She had recurrent admissions over the next 6 months with similar symptoms and noted to have a right sided SP on each occasion. Her symptoms always started between day 1 and 3 of her menstrual cycle. As medical management was unsuccessful she underwent VATS pleurectomy and right apical bullectomy in 2002. Following this intervention she had 2 further episodes of right sided pneumothoraces, one of which required an intercostal drain insertion. In view of the recurrence of the pneumothoraces, after liaison with the gynaecology team,

she was commenced on Zoladex LA injections in 2003. However her symptoms persisted and in view of the limitations in the licence for the long term use of Zoladex, she was offered TAH and BSO, which she underwent in 2005. Histology from the ovaries confirmed endometriosis. To date she has had no recurrence of her symptoms.

P575**Rituximab-induced lung disease. Case and review**

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Recent reports suggest rituximab induces pulmonary toxicity. We report a case of rituximab-induced pneumonitis in a 60 year-old woman with pemphigus, a disease which does not involve the lung. Any neoplastic disease was spread. Patient was admitted for dyspnea of 10 day-duration. Pemphigus was diagnosed 5 years earlier. She had received azathioprine (stopped 2 months before because of disease progression) thalidomide and mycophenolate mofetil without an effect. Rituximab 375 mg/m² and corticosteroids (1mg/kg) were given as rescue with a great effect, because she was finally in remission of her pemphigus vulgaris. Rituximab was stopped 1 month earlier. At admission, a dry cough and fatigue were present. There was no fever. Physical examination showed crackles bilaterally. Imaging revealed bilateral infiltrates corresponding to ground-glass on HRCT. TLC was 78% pred, CO diffusion was normal. A BAL showed 34% lymphocytes. An infectious workup was negative. Rituximab being the only drug capable of causing lung disease in our patient, was withdrawn. Steroids were kept unchanged. She improved clinically, and imaging and physiology normalized in 4 weeks. Sixteen patients of pulmonary infiltrates complicating treatments with rituximab have been reported, mainly in patients with malignant lymphomas and Hodgkin's disease. To our knowledge, the present case is the first occurring during treatment of a dermatologic condition, which simplifies causality assessment via the Naranjo score. Pathogenesis is unclear, although some cases may represent tumor lysis or cytokine release, rather than direct toxicity. Prognosis is variable. Mortality rate = 37.5%. Discontinuing rituximab is advised. The merits of corticosteroid therapy is unclear. Literature is reviewed exhaustively.