The Egyptian Journal of Hospital Medicine (Sep. 2010) Vol., 40: 435 - 441

Case Repert (Polymyositis and Interstitial Lung Disease associated with renal involvement)

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Abstract

Polymyositis with interstitial lung disease, renal involvement and positive Anti-Jo-1 antibodies in a 39-year-old male is reported. He presented with cough, dyspnoea and proximal muscle weakness. Chest imaging by X-ray and CT-scan showed wide spread reticulo-nodular densities. Urine analysis revealed RBCs Casts and albumiuria. Open lung biopsy revealed Non-specific interstitial pneumonia and muscle biopsy confirmed the diagnosis of polymyositis. Treating the patient with Prednisolone and Azathioprine showed dramatic improvement.

Introduction

Polymyositis (PM) is an idiopathic inflammatory myopathy with symmetric proximal muscle weakness, elevated skeletal muscle enzyme levels, and characteristic electromyography (EMG) and muscle biopsy findings.The reported incidence of ILD in polymyositis/dermatomyositis varies between 5% and 46% in earlier cross-sectional studies, depending on whether clinical, radiologic, functional, or pathologic criteria have been used. Renal manifestations include intrinsic renal disease which is very rare. Occasionally, severe rhabdomyolysis with myoglobinuria can result in acute tubular necrosis.

39yr old male patient smoker 20 pack year with no previous medical illnesses, hospitalized in a peripheral hospital complaining of cough, dyspnoea and low grade fever diagnosed as a case of pneumonia given intravenous antibiotic and discharged after one week. Three weeks later patient was admitted to our hospital complaining of cough dyspnoea and lower limb weakness. His physical examination showed diffuse bilateral end inspiratory crackles on chest examination and proximal muscle weakness of the lower limbs and lower limbs oedema.

His chest X- ray showed bilateral lung infiltrates, and his laboratory data shown in *(table 1)*.

Table 1

WBC	**13.500**
PCV	**38**
Platelets	**355000**
KFTs **Normal**	
Ca. **Normal**	
Phos. **Normal**	
Serum Alb. **g/L**	**28**
AST **U/L**	**922**
ALT **U/L**	**205**
LDH **U/L**	**2141**
CPK **U/L**	**9881**
ANA , ANCA , RF **ve**	**–**
Anti- jo-1	**+ ve**
Abd. U/S **Normal**	
Cardiac echo **Normal**	
PFT **Restrictive Pattern**	
DLco **50%**	
ABGs **Hypoxemia**	
hematuria with RBCs	**Urinalysis** **casts and abuminuria** **800mg/24 hours**

High resolution chest CT-scan was done **(picture1)** showed bilateral lung infiltrates with reticulation. Video assist thoracoscopic lung biopsy was done revealed Non specific interstitial pneumonia **(picture 2)**. Muscle biopsy was performed which confirm the diagnosis of polymyositis Patient started on Prednisolone 1mg/kg then tapering dose and Azathioprine ,he expressed significant improvement clinically and radiologically, his laboratory test three months after treatment shown in (table 2) and high resolution chest CT-scan 3months after treatment **(picture 3)** and 6month after treatment **(picture 4)**

Table 2

Dlco	**75 %**
PFT	**Normal**
CBC	**Normal**
KFTs	**Normal**
Serum Albumin	**35 g/L**
AST	**NORMAL**
ALT	**NORMAL**
LDH	**NORMAL**
CPK	**NORMAL**
fasting blood sugar	**182**
Urinalysis	**NIL RBCs, NIL ALBUMIN,+VE SUGAR**

Discussion

Polymyositis (PM**)** is an idiopathic inflammatory myopathy with symmetric proximal muscle weakness, elevated skeletal muscle enzyme levels, and characteristic electromyography (EMG) and muscle biopsy findings. Although the initial inciting agent remains unknown, possibilities include virus-mediated muscle injury or microvascular insult leading to release of muscle autoantigens. These autoantigens are then presented to T lymphocytes by macrophages in the muscle. Activated T lymphocytes proliferate and release cytokines such as interferon gamma (IFN-gamma) and interleukin 2 (IL-2). IFN-gamma promotes further macrophage activation and release of mediators of inflammation such as IL-1 and tumor necrosis factor-alpha (TNF-alpha). Interstitial lung disease (ILD) is a heterogeneous group of non-infectious, non-malignant disorders of the lower respiratory tract, characterized by infiltration of inflammatory cells and interstitial fibrosis. Since the original description of ILD in a case of dermatomyositis by Mills and Mathews[1] in 1956, the association of ILD with polymyositis/dermatomyositis has been widely accepted.

Renal involvement in patients with polymyositis (PM)/dermatomyositis (DM) is previously thought to be uncommon, but two main types of renal lesion have been described. First, acute tubular necrosis with renal failure related to myoglobulinemia and myoglobulinuria is a well-recognised feature of acute rhabdomyolysis. Second, chronic glomerulonephritis has been infrequently reported in a small group of patients with PM/DM

The clinical manifestations of ILD in patients with polymyositis or dermatomyositis may vary from asymptomatic to severe, rapidly progressive dyspnoea with pulmonary insufficiency and fatal outcome.

Not only the myositis itself but also the immunosuppressive treatment and secondary infections caused by immunosuppressive therapy used in these patients might lead to development of interstitial pneumonia and cause diagnostic and therapeutic dilemma.

Cough and dyspnoea are the most commonly reported symptoms, although ILD is also reported to occur in patients without any clinical overt signs of pulmonary involvement.

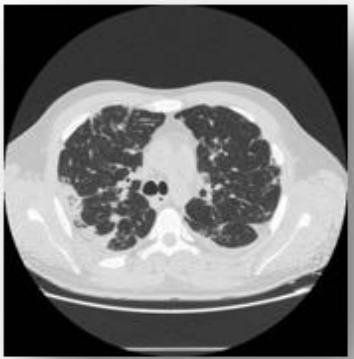
Other clinical or laboratory signs should also raise the awareness of a concomitant ILD in patients with polymyositis or dermatomyositis. The strongest predictive factor for ILD in patients with myositis is the presence of positive anti-aminoacyl t-RNA synthetase antibodies, of which the anti-histidyl t-RNA synthetase antibody (anti-Jo-1) is the most frequently found. The reported frequency of ILD in patients with anti- Jo-1 antibodies is more than 70%

It has been established that ILD can appear concomitantly with, before, or after the onset of skin or muscle manifestations**.**
Clinical respiratory symptoms are not reliable signs to detect ILD in patients with myositis, because neither cough nor dyspnoea may be present as early signs.

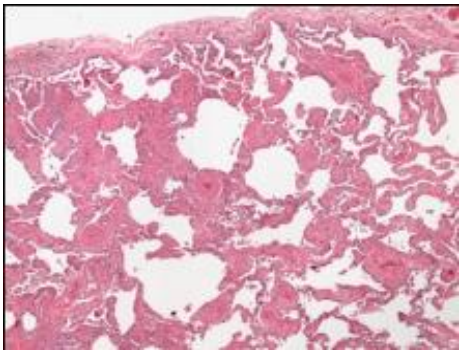
The most useful tests to diagnose ILD are pulmonary function tests, which typically show a restrictive ventilatory defect with decreased total lung capacity, functional residual capacity, residual volume, forced expiatory volume in 1 second (FEV_1), and forced vital capacity (FVC), but with a normal or elevated FEV_1/FVC ratio and a decreased diffusing capacity for carbon monoxide (DLco) Not all of these abnormalities may be found in every patient, however. The most sensitive test seems to be the DLco.
HRCT is now widely used not only for detection of ILD but also for identifying the extent and severity of the disease as well as to discriminate between fibrotic Lung disease and active inflammation in the lungs lung biopsy.
Lung biopsies are not routinely performed in patients with myositis with signs of ILD for diagnostic purposes because of the potential morbidity associated with surgical lung biopsy.

Transbronchial biopsies are rarely helpful in the diagnostic procedures, although they are usually abnormal, because the histopathologic findings are nonspecific

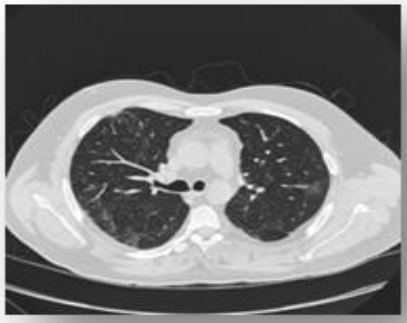
Anti-Jo-1 antibodies could not be used as a diagnostic tool for ILD, but because these auto-antibodies are highly associated with ILD, their presence requires careful evaluation of lung involvement using lung function tests and HRCT
Non specific interstitial pneumonia followed by organizing pneumonia was the most commonly observed histologic pattern inpatient with myositis with ILD**.**
Patients with BOOP and cellular NSIP tend to have the best prognosis and response to corticosteroid treatment, whereas those with DAD have the worst prognosis. Patients with UIP tend to have an intermediate course.
The optimal treatment for myositis-associated ILD is not known. No published controlled trials exist on the effects of different therapies in polymyositis/dermatomyositis with associated ILD. Thus, available information on treatment efficacy is based on small case series or case reports.
Corticosteroid treatment as a single agent is often not sufficient to cause improvement of ILD. Furthermore, the high doses required over a long period are often associated with severe side effects. Thus, other immunosuppressive agents are often required. The most frequently used drugs with reported beneficial effects on lung function are Cyclophosphamide, Cyclosporine A, Azathioprine, and Methotrexate. The response rate may be higher when treatment is initiated early in the course of the disease, before irreversible changes have developed.



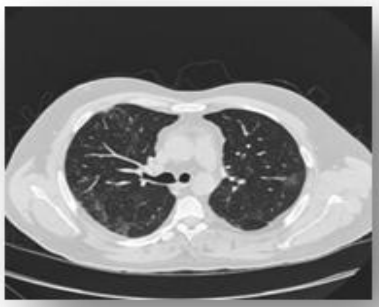
PICTURE 1: showed bilateral lung infiltrates with reticulation



PICTURE 2: Focal areas of diffuse alveolar septal fibrosis extending to the pleural surface were seen - nonspecific interstitial pneumonia.



PICTURE 3: chest CT-scan 3months after treatment. Shows significant improvement with reduce in lung infiltrates and reticulo-nodular shadows



PICTURE 4: 6 months after treatment

ABBREVIATIONS:

ALT	**Alanine aminotransferase**
ANA	**Antinuclear antibodies**
ANCA	**Antineutrophil cytoplasmic antibodies**
AST	**Aspartate aminotransferase**
BOOP	**Bronchiolitis obliterans organizing pneumonia**
DAD	**Diffuse alveolar damage**
DLCO	**Diffusing capacity for carbon monoxide**
DM	**Dermatomyositis**
EMG	**Electromyography**
FEV1	**Forced expiratory volume in 1 second**
FVC	**Forced volume capacity**
HRCT	**High resolution computed tomography**
ILD	**Interstitial lung disease**
KFT	**Kidney function test**
NSIP	**Non specific interstitial pneumonia**
PCV	**Packed cell volume**
PFT	**Pulmonary function test**
RF	**Rheumatoid factor**
RNA	**Ribonucleic acid**
UIP	**Usual interstitial pneumonia**

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التهاب العضلات المتعدد وأمراض الرئة الخلالي المرتبطة بالمشاركة الكلويه

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الالتهاب العضلي المتعدد: هو من امراض المناعة الذاتية الذي يؤدي الى حدوث التهاب في العضلات وحدوث ضعف فيها. أعراض المرض متفاوتة حيث يسبب اعياء وضعف عضلي شديد في العضلات المحوريه كعضلات الكتفين والحوض, الام في المفاصل, صعوبه في البلع, صعوبة في التنفس نتيجه اصابه عضلات الصدر. يصاحب المرض اصابات رئويه تتراوح بين 5% – 46% مثل ذات الرئه الخلاليه الغير اعتياديه وفشل الجهاز التنفسي بالاضافه الى اصابه الكليتين النادره الحدوث على شكل بيله الغلوبين العضلي ونخر انبوبي كلوي حاد. يشخص بعد الفحص الاكلينيكي ومن ثم قياس نشاط العضلات عن طريق جهاز رسم العضلات الكهربائي, ارتفاع نسبه انزيمات العضلات.

ويعتبر اخذ خزعه من النسيج العضلي وفحصها تحت المجهر الاختيار الافضل لتشخيص المرض.

دراستنا كانت على مريض 39 عام, مدخن يشكو من سعال وضيق في التنفس بالاضافه الى ضعف عضلي سفلي حاد, وبعد الفحوصات المخبريه والشعاعيه تبين وجود ارتشاح رئوي التهابي وارتفاع الانزيمات العضليه ووجود قوالب كرويه حمراء وزلال بولي. تم عمل خزعه عضليه دلت على وجود الالتهاب العضلي المتعدد ووجود ذات الرئه الخلاليه الغير اعتياديه.

المريض عولج بالكورتيكوستيرويدات مع استجابه ملحوظه للعلاج.

الدراسه تدل على مدى اصابة الرئه والكليتين عند مرضى الالتهاب العضلي المتعدد و استجابتهم للعلاج.