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Dermatomyositis Masquerading as Pulmonary Embolism

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Abstract
A 61-year-old Caucasian was admitted to Department of Chest Diseases and Tuberculosis, Medical University of Bialystok, Poland for progressive muscle weakness and weight loss. Eighteen months prior to admission, the patient had been diagnosed with pulmonary embolism. At that point he was started on Enoxaparin QD. Past medical history was unremarkable. In the interim, the patient developed fever, myalgia and progressive dyspnea. Physical examination on admission revealed a rash on his upper torso and back, and the extensor surfaces of all four extremities. Laboratory values included CPK 8229, MB fraction 219, LDH 981. Chest X-ray and CT scan revealed bilateral patchy consolidations and ground-glass opacities. EMG was consistent with myositis. The patient was started on solumedrol 40 mg i.v., b.i.d., and then switched to prednisone 40 mg b.i.d. His symptoms and muscle strength improved remarkably. The patient was discharged with prednisone with an outpatient follow up.

Key words: dermatomyositis, polymyositis, pulmonary embolism

Introduction

Dermatomyositis (DM) is an idiopathic inflammatory myopathy with characteristic skin manifestations [1, 2]. The prevalence of the disease is 1-10 cases per million in adults and 1-3.2 cases per million in children [3]. In 1975, Bohan and Peter [4, 5] first suggested a set of criteria to aid in the diagnosis and classification of polymyositis (PM) and DM as follows: progressive proximal symmetrical weakness, elevated levels of muscle enzymes, an abnormal finding on electromyography, an abnormal finding on muscle biopsy, and skin manifestations which are compatible with cutaneous disease (Table 1).

Case Report

The study was performed according to the standards set by the Helsinki Declaration of 1975, regarding the Human Research and was approved by an institutional Ethics Committee. Informed consent was obtained from the patient described in this article.

A 61-year-old Caucasian male was admitted to Department of Chest Diseases and Tuberculosis, Medical University of Bialystok for progressive muscle weakness. The patient also developed a rash on his upper torso and back, and the extensor surfaces of all four extremities. The patient complained of generalized muscle weakness that had been present for several months but had now progressed remarkably. One and half year prior to admission, he had been admitted to the department for recurrent hemoptysis and was diagnosed with pulmonary embolism, fitting all clinical and radiological criteria. At that point he was started on enoxaparin QD. Past medical history and surgical history were unremarkable, and the patient denied the use of any tobacco or recreational drugs. He retired 6 years prior to admission. Hemoptysis stopped after first two weeks of hyperfractionated heparin treatment. He admitted to losing about 53 kilos in 18 months prior to admission, with accompanying decrease in appetite. In the interim, he developed fever, myalgia, and progressive dyspnea. He denied any previous episodes similar to this, and reported no family history of musculoskeletal disease. Physical examination revealed an ill-appearing man with an oral temper-

Table 1. Diagnostic criteria of polymyositis and dermatomyositis, according to Bohan and Peter [4, 5].

1. Progressive symmetrical general muscle weakness
2. Typical for myositis histopathology findings
3. Elevated plasma level of CK and/or aldolase
4. EMG findings typical for primary myopathy
5. Typical skin findings (Gottron's sign, helitrope rash of eyelids, upper torso, arms)

	Number of symptoms 5		
Diagnosis	**Confirmed**	**Suggestive**	**Possible**
PM	4	3	2
DM	3-4	2	1

plus characteristic skin findings

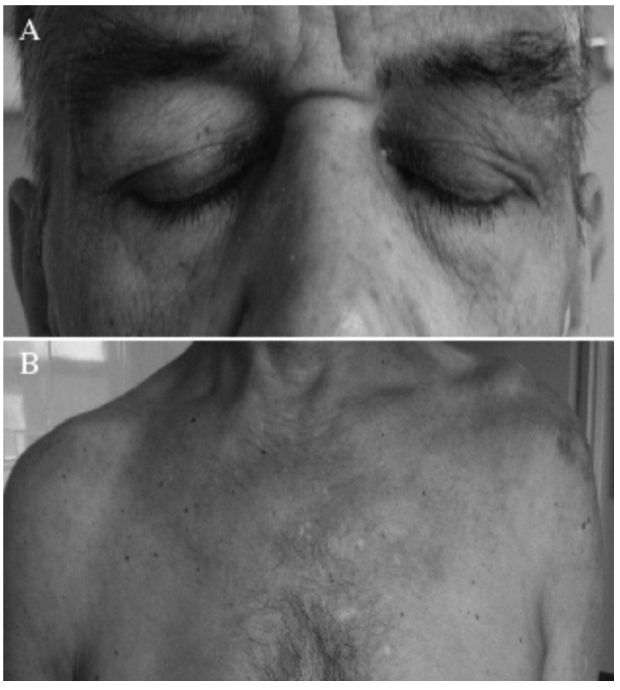

Fig. 1. Skin manifestations I. Panel A - Heliotrope rash and Panel B - Violet discoloration and swelling of the eyelids.

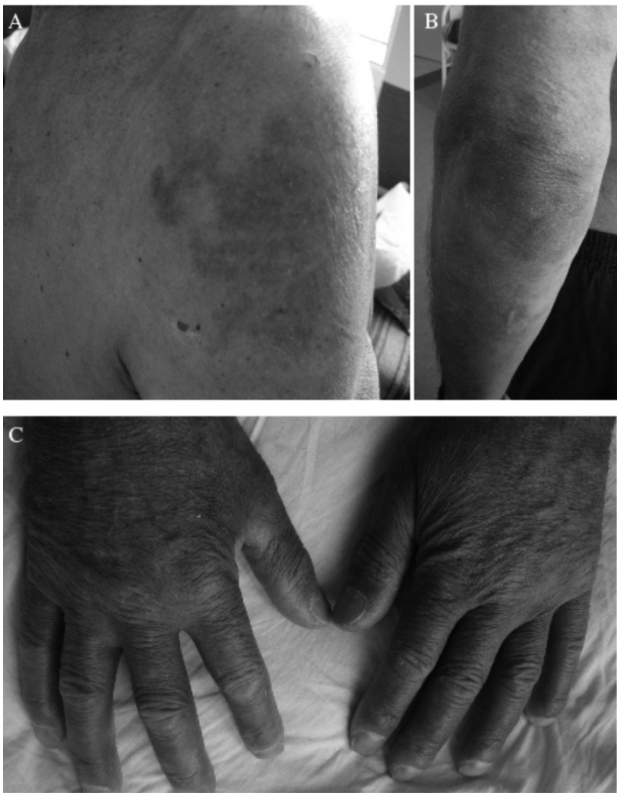

Fig. 2. Skin manifestations II. Panel A - Rash on shoulders, Panel B - Erythematous papules over joints – 'Gottron's sign', and Panel C - Hypertrophic changes of palms and fingers – 'Machanic's hands'.

ature of 38.2°C, a pulse rate of 60 beats/min, and normal blood pressure. His skin had purple confluent discoloration and swelling of the eyelids (Fig. 1A), the

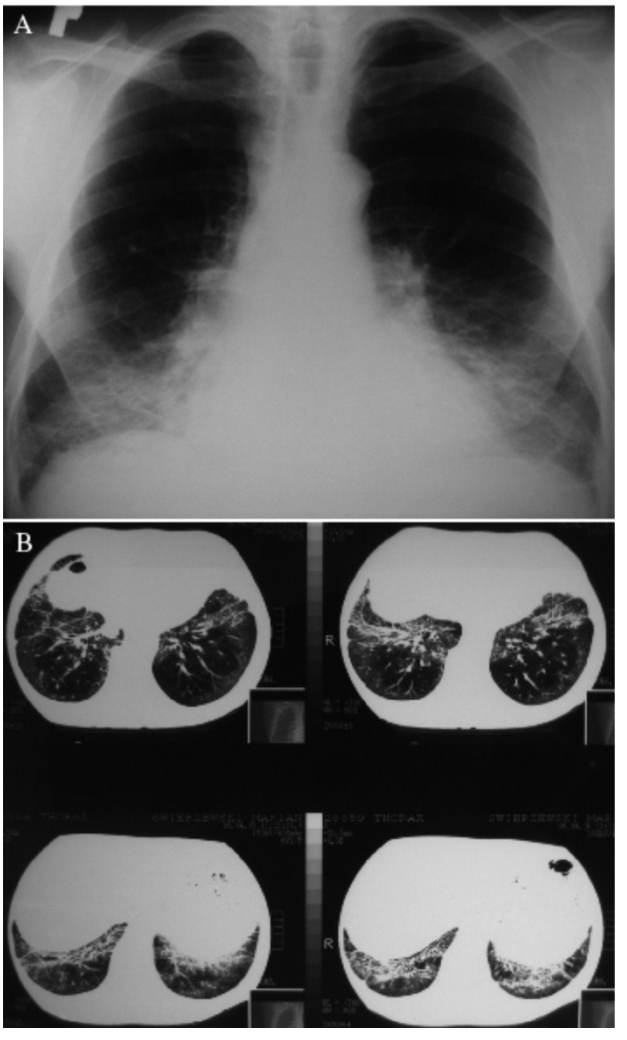

Fig. 3. X-Ray (Panel A) and HRCT (Panel B) findings. Bilateral patchy consolidations and ground-glass opacities.

V-shape rash with areas of hyperpigmentation of his upper torso (Fig. 1B). The rash was also present on his shoulders (Fig. 2A), and erythematous papules over joints (Fig. 2B). Hypertrophic changes of palms and fingers (Fig. 2C), were also found. Subtle dry cracles and wheezing were found on auscultation. Chest X-ray and CT scan revealed bilateral patchy consolidations and ground-glass opacities (Fig. 3A and B). On admission, laboratory values included CPK 8229 U/l, MB fraction 219 U/l, LDH 981 U/l, and D-dimer 0.96 ng/ml. EMG was consistent with myositis. At admission, the patient was started on solumedrol 40 mg i.v, b.i.d., and then switched to prednisone 40 mg b.i.d. After two weeks of therapy, the patient's symptoms improved remarkably. His muscle strength improved and he was able to participate in physical therapy. The patient was discharged back to his facility with prednisone with an outpatient follow up.

Discussion

This patient was diagnosed with dermatomyositis, fitting 4 out of the five diagnostic criteria, as outlined above: proximal muscle weakness, CPK and LDH ele-

vations, consistent with myositis EMG, and skin manifestations [4, 5]. The latter were as follows: purple confluent discoloration and swelling of the eyelids - heliotrope rush, the V-shape rash with areas of hyperpigmentation of his upper torso and shoulders, back, knees, and erythematous papules over joints – 'Gottron's sign', and hypertrophic changes of palms and fingers – 'Mechanic's hands'. The diagnosis is confirmed if a patient presents with at least 4 diagnostic criteria for PM or 3 for DM [4, 5]. The fifth criterion which is confirmation of inflammatory infiltrations on muscle biopsy was not done in this case. Since our patient exhibited clinical and laboratory findings consistent with inflammatory myopathy with skin manifestations present, our diagnosis was consistent with dermatomyositis.

The cause of the disease remains unknown. Altered immune response towards viral infection is considered to play a major role [1, 2]. Vascular deposits of immune complexes and complement, leading to endothelial cell injury and small vessel obstruction are most important pathogenetic factors underlying the disease [6]. The muscles and endothelial cells are infiltrated with predominance of T CD4+ cells in DM and T CD8+ cells in PM patients [6]. An association of tumor necrosis factor alpha and HLA polymorphisms with adult dermatomyositis has recently been reported [7, 8]. Our patient exhibited all reported skin manifestations of DM which were: a heliotrope rash, which is a purple discoloration and swelling of the eyelids, Gottron's sign demonstrated with erythematous papules over joints, and mechanic's hands, which are: erythematous and hypertrophic skin changes of palms and fingers [1-5]. Hemoptysis, progressive dyspnea, subtle dry cracles and wheezing on auscultation, with bilateral patchy consolidations and ground-glass opacities on chest X-ray and CT scans confirmed lung interstitium disease association in our case. Hemoptysis and pulmonary embolism, lack of significant muscle weakness at the onset of the disease masqueraded underlying DM and delayed proper diagnosis. Lung involvement has been reported in minority of PM and DM cases, influencing the course and worsening prognosis in most of the cases [9, 10]. An association with other connective tissue disorders (overlap syndrome) and malignancy has also been reported [9, 10, 11, 12, 13]. The course of the disease is usually steadily progressive, leading to profound symmetrical skeletal muscle weakness of all extremities. Patient management includes careful evaluation for underlying malignancy, physical therapy, antihistamines, sunscreen, and oral corticosteroids [14]. Alternative therapies include corticosteroid-sparing agents, cytotoxic drugs, and more recently monoclonal antibodies with cutaneous lesions treatment [14, 15, 16, 17, 18]. Irresponsive disease, delay in diagnosis and the underlying malignancies worsen disease prognosis.

Conflicts of interest: The authors had no conflicts of interest to declare in relation to this article.

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