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ORIGINAL ARTICLE

Outcome of patients having dermatomyositis admitted to the intensive care unit

Yaniv Sherer · Daniel Shepshelovich · Tamara Shalev · Yael Haviv · Eran Segal · Michael Ehrenfeld · Yair Levy · Rachel Pauzner · Yehuda Shoenfeld · Pnina Langevitz

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Abstract Patients having systemic rheumatic diseases constitute a small percentage of admissions to the medical intensive care units (ICUs). Dermatomyositis (DM) is one of the rheumatic diseases that have secondary complications that may lead to a critical illness requiring hospitalization in the ICU. Herein, we present the features, clinical course, and outcome of critically ill patients having DM who were admitted to the ICU. The medical records of six DM patients admitted to the ICU in a large tertiary hospital in a 12-year period were reviewed. The mean age of patients at time of admission to the ICU was 38 (range 16–37). Mean disease duration from diagnosis to admission to the ICU was 1.6 years (range 1 month–8 years), while the main reason for admission to the ICU was acute respiratory failure. Two of six patients died during the hospitalization. The main causes of death were respiratory complications and sepsis. The outcome of DM patients admitted to the ICU was generally not different from the outcome of other patients hospitalized in the ICU. The main reason for hospitalization was acute respiratory failure. As there are many reasons for respiratory failure in DM, an early diagnosis and aggressive appropriate treatment may help to further reduce the mortality in these patients.

Keywords Dermatomyositis · Intensive care unit · Mechanical ventilation · Mortality

Y. Sherer (✉) · D. Shepshelovich · T. Shalev · Y. Levy · Y. Shoenfeld
Department of Medicine B and Center for Autoimmune Diseases, Sheba Medical Center,
Tel Hashomer,
Ramat Gan, Israel
e-mail: sherery@017.net.il

Y. Sherer · D. Shepshelovich · T. Shalev · M. Ehrenfeld · Y. Levy · R. Pauzner · Y. Shoenfeld · P. Langevitz
Sackler Faculty of Medicine, Tel-Aviv University,
Tel-Aviv, Israel

Y. Haviv · E. Segal
Intensive Care Unit, Sheba Medical Center,
Tel Hashomer, Israel

M. Ehrenfeld · R. Pauzner · P. Langevitz
Rheumatology Unit, Sheba Medical Center,
Tel Hashomer,
Ramat Gan, Israel

Y. Shoenfeld
Laura Schwarz-Kipp Research of Autoimmune Diseases,
Tel-Aviv University,
Tel-Aviv, Israel

Introduction

Patients having systemic rheumatic disease constitute only a small group of all intensive care unit (ICU) admissions [1–4]. Life-threatening illnesses requiring ICU hospitalization are likely to develop in these patients because of exacerbation of the underlying disease, development of new manifestations of the rheumatic disease, infections, adverse effects of drugs, or acute serious illness unrelated to the systemic rheumatic disease but whose manifestations are altered or exacerbated by it. In most of the patients, more than one of these factors coexist, and the complexity of these conditions cause the critical illness [1]. More than 50% of the admissions of these patients to the ICU are due to infections, and 25–35% are the result of an exacerbation of the rheumatologic disorder [1]. Dermatomyositis (DM) is one of the rheumatic diseases that have secondary complications that may lead to a critical illness requiring hospitalization in the ICU. DM is an idiopathic inflamma-

tory disorder that usually presents with progressive symmetrical muscle weakness over several months, but an acute presentation is occasionally seen [5]. Shoulder and pelvic girdle muscles are usually the major muscle groups affected in DM. Although facial muscles are spared, neck flexion muscles are weakened in up to 50% of patients. Pharyngeal muscle involvement may present with dysphonia or dysphagia. Myalgias and muscle tenderness are less common than in other inflammatory myopathies, such as infection and drug-related myositis, but may occur in up to 50% of patients [6].

Cardiac and respiratory complications of DM are the main concerns for an intensivist. Cardiac involvement presents with tachyarrhythmias, conduction abnormalities, or heart failure with a dilated cardiomyopathy. Chronic pulmonary hypertension may result from several mechanisms, including underlying cardiomyopathy, hypoxia associated with either chronic hypercapnic respiratory failure or interstitial lung disease, or the development of a primary pulmonary hypertension-like syndrome [7]. Respiratory complications with DM include abnormal central respiratory drive, pharyngeal muscle impairment, respiratory muscle weakness, interstitial disease, pneumonia caused by aspiration or therapy-related immunosuppression, and drug-induced lung disease [7]. Respiratory muscle dysfunction with inspiratory and expiratory muscle involvement has been reported in up to one third of patients. An analysis of 70 patients with DM and diffuse interstitial lung disease revealed an initial musculoskeletal presentation (myalgias, arthralgias, weakness) in 25 patients (36%), pulmonary presentation in 21 patients (30%), and a combination of musculoskeletal and pulmonary symptoms in 15 patients (21%) [8]. Herein, we present the features, clinical course, and outcome of critically ill patients having DM who were admitted to the ICU.

Materials and methods

The study was conducted in the ICU of Sheba Medical Center, in Tel-Hashomer, Israel. All available information about clinical and laboratory features in DM patients that were hospitalized in the ICU in the period 1991–2002 was retrospectively analyzed. All patients were diagnosed according to the criteria described in [9]; disease duration was determined from the onset of the first symptoms. The following information was obtained for every patient: age, sex, and duration of DM, manifestation of the disease, length of hospital stay before admission to the ICU, length of stay in the ICU, the indication for the admission to ICU, and whether it was an emergency or an elective admission. Clinical, hematological, and serological characteristics and previous treatment regimens were recorded for each patient. The Acute Physiology and Chronic Health Evaluation (APACHE II) score was used to determine the severity of illness in the first 24 h on admission to the ICU. APACHE II scores were calculated from the ICU charts and records retrospectively. The management during the patient hospitalization in the ICU was recorded, including the requirement for mechanical ventilation, hemodialysis, ionotripis support, and the duration of their use. Special attention was given to the use of various treatment regimes before the admission to the ICU and thereafter. The causes of death were determined on the basis of clinical data, and, where available, postmortem examination was reviewed.

Results

Patient characteristics

A total of six patients having DM that were hospitalized in the ICU in a 12-year period were included in the study. In the same period, 42 other patients having DM or polymyositis were treated in the hospital and were not referred to the ICU. All six were women. Mean age at the time of admission was 38 (range 18–67). Mean disease duration from diagnosis to admission to the ICU was 1.6 years (range 1 month–8 years). The major clinical characteristics of the patients are shown in Table 1. Five of the six patients had symptoms for less than a year; four had symptoms for less than 6 months. Three of the patients were diagnosed during their hospital admission and were not treated earlier. Only one patient was diagnosed as having DM for more then 1 year before the admission and was treated with a steroid-sparing agent (methotrexate). In that period of time, six patients having amyotrophic lateral sclerosis and ten having Guillain–Barre syndrome were followed in the ICU.

ICU course and outcome

The reasons for admission to the hospital and the ICU varied among patients (Table 2). Three patients were admitted to the hospital because of respiratory failure, two

Table 1 Clinical characteristics of dermatomyositis patients before admission

Patient no.	Age/ sex	Disease duration	Duration of treatment with steroids	Other chronic treatment regimes
1	67/F	1.5 months	0 years	No
2	18/F	1 month	0 years	No
3	40/F	8 years	8 years	Methotrexate
4	55/F	2 months	0 years	No
5	30/F	12 months	1 year	No
6	18/F	6 months	4 months	No

Table 2 Course and outcome of dermatomyositis patients admitted to the ICU

Patient no.	Age/Sex	Reason for admission to the hospital	Reason for admission to the ICU	Length of stay in the hospital (days)	Length of stay in the ICU (days)	APACHE II on admission to ICU	Outcome
1	67/F	Respiratory failure	Respiratory failure	27	18	18	Death because of respiratory failure
2	18/F	Respiratory failure	Respiratory failure	130	14	16	Discharge
3	40/F	Respiratory failure	Respiratory failure	20	1	18	Discharge
4	55/F	Peritonitis	Peritonitis	35	1	20	Discharge
5	30/F	Respiratory failure, dysphagia	Respiratory failure	360	60	27	Death because of severe sepsis
6	18/F	Acute respiratory failure, dysphagia,	Respiratory failure	285	16	22	Discharge

were admitted because of respiratory failure and dysphagia, and one patient was admitted because of acute peritonitis. The mean duration of treatment in the ICU was 19 days (range 1–60). The mean duration of hospitalization was 162 days (range 27–360). The mean APACHE II score on admission to the ICU was 20.16 (range 16–27). Patients 1–3 were admitted because of respiratory failure. Patient 1 died in the ICU because of respiratory failure secondary to a severe interstitial lung disease, patient 2 was discharged from the hospital after 130 days, and patient 3 was discharged after 20 days. Patient 4 was admitted because of acute peritonitis after acute bowel obstruction. He was admitted to the ICU after an emergency operation and was discharged from the hospital 35 days later. Patient 5 was admitted to the hospital because of severe dysphagia and acute respiratory failure. She was transferred to the ICU because of severe sepsis, probably because of recurrent aspirations. She died in the ICU. Patient 6 was admitted because of dysphagia, respiratory failure, and sepsis. She was discharged from the hospital 3 weeks later.

The ICU management and treatment of the patients is shown in Table 3. Five of the six patients received mechanical ventilation, four of them for prolonged periods of time (mean 35 days, range 18–60). Three of the six had systemic infections and were treated by antibiotics. One of them had an intraabdominal infection, and the other two suffered from *Pseudomonas aeuroginosa* sepsis. Five of the six were treated by steroids during their ICU admissions. None of the patients required hemodialysis during their stay in the ICU, and none had arrhythmias or any sign of heart failure.

Discussion

The overall mortality rate of individuals with DM is about four times that of the general population; death is usually due to pulmonary, renal, and cardiac complications. Women, blacks, and those severely affected at presentation or treated after long delays have worse prognosis. Unfavorable outcome is also seen in patients with significant dysphagia, associated cancer or connective tissue disease, and serum antibodies to Jo-1 and SRP. Evidence from several series suggests that patients seen at tertiary referral centers may have less-favorable outcome when compared with patients seen at smaller community hospitals, probably

Table 3 Management during ICU hospitalization

Patient no.	Dialysis	Mechanical Ventilation	Length of mechanical ventilation (days)	Microbiology	Antimicrobial treatment	Steroid treatment
1	No	Yes	18	No	No	Yes
2	No	Yes	30	No	No	Yes
3	No	No	0	No	No	Yes
4	No	Yes	1	Abdominal cultures—*E. Coli*	Yes	No
5	No	Yes	60	Blood cultures—Pseudomonas	Yes	Yes
6	No	Yes	30	Blood cultures—Pseudomonas	Yes	Yes

because they represent a population with more severe disease that is less responsive to therapy [10, 11]. In our study, the overall mortality was 33.3% for patients with DM admitted to the ICU, equal to the overall mortality there (30–40% in our ICU). Of special note are the patients' young age (mean 38, range 18–67, two of them 18 years old) and the short lag of time from diagnosis to a severe life-threatening disease. Three out of six patients had DM for less then 2 months and five out of six for less then 1 year. Two of the patients (patients 1–2) were diagnosed with DM in the ICU. Although it is known that DM patients might have an acute presentation [5], most of the patients in our study deteriorated right after the diagnosis, before proper treatment could be initiated. Only three out of the six patients were treated at all before their admissions, one of them only for a few months, and only one of them was treated by a steroid-sparing drug. Whether this indicates that the disease is more severe initially or that well-treated patients have a more stable disease and less risk for an acute exacerbation is yet unknown.

Five of the six patients were admitted to the ICU because of respiratory failure. Two of them experienced severe dysphagia. Pulmonary complications are a common cause of death in patients with DM. DM patients are prone to develop chronic pulmonary hypertension, abnormal central respiratory drive, pharyngeal muscle impairment leading to recurrent aspirations and pneumonia, respiratory muscle weakness, interstitial lung disease, pulmonary infections because of therapy-related immunosuppression, and drug-induced lung disease [7]. Numerous investigators reported various respiratory pathologies in patients with DM [12–15]. It is not uncommon for one patient to suffer from several pulmonary pathologies at the same time, a phenomena that aggravates one's condition. In our study, patient 1, who was the oldest patient (67 years old), suffered from severe interstitial lung disease and progressive muscle weakness. She died from respiratory failure. Patient 5 had severe dysphagia, a major risk factor for DM patients, and interstitial lung disease. She died from severe sepsis complicated by respiratory failure.

Treatment for severe respiratory failure in DM patients requires accurate diagnosis and aggressive treatment. Interstitial lung disease exacerbation should be treated with corticosteroids. Respiratory muscle weakness and hypercapnea should be treated by mechanical ventilation and corticosteroids. Aspiration pneumonia and sepsis is treated with broad-spectrum antibiotics and ionotropic drugs as necessary; dysphagia usually responds to a course of corticosteroids, whereas surgical treatment is rarely indicated. Five of the six patients in our study were treated with corticosteroids for respiratory failure. When treatment with corticosteroids fails, the patient should be treated with agents such as methotrexate, azathioprine, cyclophosphamide, intravenous immune globulin, cyclosporine, tacrolimus, alkylating agents, and tumor necrosis factor inhibitors [16–19]. Patients who do not respond to immunosuppressive therapy could be treated by mechanical ventilation at home.

In conclusion, the outcome for DM patients requiring admission to the ICU was not different than the average ICU outcome. The most common indication for admission was respiratory failure because of exacerbation of muscle weakness, interstitial lung disease or recurrent aspirations, pneumonia, and sepsis. Most of the DM patients who required ICU treatment were diagnosed with DM in the months preceding the hospitalization. Therefore, we suggest that patients who are newly diagnosed with DM should be more closely monitored, at least until corticosteroid treatment has been prescribed. However, limitations in the study size and design do not allow us to draw a definitive conclusion regarding the outcome of DM patients admitted to the ICU and to identify subgroups of these patients with a worse prognosis.

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References

1. Janssen NM, Karnad DR, Guntupalli KK (2002) Rheumatologic diseases in the intensive care unit: epidemiology, clinical approach, management and outcome. Crit Care Clin 18:720–748
2. Kollef MH, Enzenauer RJ (1992) Predicting outcome from intensive care for patients with rheumatologic diseases. J Rheumatol 19:1260–1262
3. Godwau B, Mortier E, Roy PM et al (1997) Short and long-term outcomes for patients with rheumatic diseases admitted to the intensive care unit: a prognostic study of 181 patients. J Rheumatol 24:1317–1323
4. Shalev T, Haviv Y, Segal E, Ehrenfeld M, Pauzner R, Levy Y, Langevitz P, Shoenfeld Y (2006) Outcome of patients with scleroderma admitted to intensive care unit. A report of nine cases. Clin Exp Rheumatol 24:380–386
5. Dalakas MC (1991) Polymyositis, DM, and inclusion-body myositis. N Engl J Med 325:1487–1498
6. Tymms KE, Webb J (1985) Dermatopolymyositis and other connective tissue diseases. A review of 105 cases. J Rheumatol 12:1140–1148
7. Schwartz M (1998) The lung in polymyositis. Clin Chest Med 19:701–712
8. Douglas WW, Tazelaar HD, Hartman TE et al (2001) Polymyositis-DM associated interstitial lung disease. Am J Respir Crit Care Med 164:1182–1185
9. Bohan A, Peter JB (1975) Polymyositis and dermatomyositis. N Engl J Med 292:344–347
10. Astudillo LM, Arriero M, Sailler L, Dingremont CF, Arlet PM (2001) Hypercapnic coma due to diaphragmatic involvement in a patient with DM. Clin Exp Rheumatol 19(4):456–458, Jul–Aug
11. Kuwana M (2004) Collagen diseases with pulmonary involvement. Nihon Rinsho Men'eki Gakkai Kaishi 27(3):118–126, Jun
12. Sakamoto N, Mukae H, Fuji T, Yoshioka S, Kakugawa T, Yamaguchi H, Hayashi T, Kohno S (2004) Nonspecific interstitial

pneumonia with poor prognosis associated with amyopathic DM. Intern Med 43(9):838–842, Sep
13. Cherin P, Herson S (1994) Indications for intravenous gamma-globulin therapy in inflammatory myopathies. J Neurol Neurosurg Psychiatry 57:50–54
14. Dalakas MC et al (1993) A controlled trial of high dose intravenous immune globulin infusions as treatment for DM. N Eng J Med 329:1993–2000
15. Dalakas MC (1994) How to diagnose and treat the inflammatory myopathies. Semin Neurol 14:137–145
16. Joffe MM et al (1993) Drug therapy of the idiopathic myopathies: Predictors of response to prednisone, azathioprine and methotrexate and a comparison of their efficacy. Am J Med 94:379–387
17. Haskard DO (1983) Successful treatment of DM complicated by ventilatory failure. Ann Rheum Dis 42(4):460–461, Aug
18. Shinohara T, Hidaka T, Matsuki Y, Ishizuka T, Takamizawa M, Kawakami M, Kikuma H, Suzuki K, Nakamura H (1997) Rapidly progressive lung disease associated with DM responding to intravenous cyclophosphamide pulse therapy. Intern Med 36 (7):519–523, Jul
19. Selva-O'Callaghan A, Sanchez-Sitjes L, Munoz-Gall X, Mijares-Boeckh-Behrens T, Solans-Laque R, Angel Bosch-Gil J, Morell-Brotad F, Vilardell-Tarres M (2000) Respiratory failure due to muscle weakness in inflammatory myopathies: maintenance therapy with home mechanical ventilation. Rheumatology (Oxford) 39(8):914–916, Aug