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Brief Report

Plasma-exchange as a "rescue therapy" for dermato/polymyositis in acute phase. Experience in three young patients

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ABSTRACT

There are few data in the literature supporting the efficacy of plasma-exchange in dermato/polymyositis. The authors report three cases of patients with acute disease phase showing severe pharyngo-esophageal muscle weakness unresponsive to conventional therapy (corticosteroids and immunosuppressant agents) who were treated with plasma-exchange. As the patients were at high risk of "aspiration pneumonia", tracheostomy and PEG tubes were placed.

The patients underwent a series of plasma-exchange for a mean of 15 weeks, during which time they progressively recovered muscle strength, their serum muscle enzyme values returned to normal levels, and MRI showed resolution of muscle edema. The tracheostomy and PEG tubes could be removed.

Our findings suggest that plasma-exchange in association with immunosuppressant agents could play a relevant role in the management of dermato/polymyositis in acute phase.

1. Introduction

Dermatomyositis (DM) and polymyositis (PM), which are the main idiopathic inflammatory myopathies [1], are commonly characterized by: a) subacute onset; b) proximal, symmetric muscle weakness and fatigue; and c) mononuclear cell infiltration in muscle tissue. Although DM and PM share common clinical features, skin manifestations (heliotrope rash and Gottron's papules), which often precede or accompany muscle weakness, are peculiar to the former [2].

DM/PM is a rare disease, more common in black race. The exact prevalence and incidence are unknown. Prevalence data for DM/PM vary from 5 to 11 cases per 100 000 inhabitants. The reported incidence ranges from 1.2 to 17 new cases/million population/year. DM/PM has been observed at any age, although PM usually affects adults. DM/PM in adults affects women twice as often as men [3].

Despite aggressive treatments (high dose prednisone and immunosuppressant agents), which are especially important during acute phases to avoid severe complications such as respiratory failure, DM/PM shows poor outcome in some patients.

Therapeutic plasma exchange (TPE) has been used since the 1980s to manage connective tissue diseases [4]. The procedure's immunomodulatory effects are probably linked to the removal of circulating autoantibodies, immune complexes, cytokines and other mediators involved in the disease's pathogenesis. Over recent decades, the Rheumatology Unit of the Padova University, in collaboration with

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the Apheresis Unit of the Padova Hospital, has developed exhaustive experience in treating connective tissue diseases with TPE [5]. Here we report on three young DM/PM patients who were treated with TPE as a "rescue therapy" measure during acute disease phases when traditional therapies failed.

2. Case reports

Patient A: a 17-year-old male was transferred to the Rheumatology Unit from a peripheral Hospital one month after acute onset of DM/PM. The patient's clinical picture at admission, including the results of diagnostic tests, is outlined in Table 1.

The patient was treated with high doses of prednisone (1 g iv for 3 days and then 1 mg/kg/day orally) together with methotrexate (MTX, 20 mg/wk im) without any clinical benefits for 4 weeks [manual muscle testing (MMT)-8 = 31, normal range 130–150], although there was a reduction in serum creatine kinase (CK) and myoglobin (Mb) levels (Table 2). At the same time, recurrent episodes of aspiration of salivary secretions led us to ask for a consultation with surgeons for tracheostomy and percutaneous endoscopic gastrostomy (PEG) tube placement. High-dose human intravenous immunoglobulin (IVIG) therapy was concomitantly added to the patient's therapy regimen, but since he did not tolerate the first infusion, it was suspended. A collegial decision was then made to associate TPE to prednisone and MTX.

TPE was carried out following a schedule adopted by our Apheresis Unit to treat immune-mediated systemic diseases [6]. The protocol utilized was a modified version of one used by French Haemapheresis Centers [7]. We modified their protocol scheduling the sessions more frequently in view of obtaining a more rapid therapeutic effect. The sessions were carried out twice a week during the first month, then once a week until TPE was withdrawn. TPE was performed using a COBE Spectra continuous blood cell flow separator (Terumo BCT, Lakewood, CO, USA). Seventy to 100% of patient's plasma volume was exchanged at each session and the replacement fluid was a mixture of 70% human albumin (4%) and 30% saline. Anticoagulation was ensured by using Acid Citrate Dextrose Formula A anticoagulant in a 1:12/1:15 ratio.

Table 1
Clinical picture at admission to Rheumatology Unit and the results of diagnostic tests in three patients with DM/PM.

	Patient A	Patient B	Patient C
Skin manifestations	Periorbital heliotrope rash Gottron's sign on the hands	Periorbital heliotrope rash Gottron's papules on the hands	Facial erythematous rash Gottron's papules on the hands
Muscle symptoms	Weakness of proximal muscles of upper and lower limbs Severe dysphagia	Weakness of proximal muscles of lower limbs. Fatigue Dysphagia	Upper and lower limb weakness Dysphagia
Manual muscle test – 8 (normal range: 130–150)	25	31	26
Serum creatine kinase (normal range: 20–180 U/L)	2637 U/L	4080 U/L	3041 U/L
Serum myoglobin (normal range: 12–70 μg/L)	1089 μg/L	844 μg/L	734 μg/L
Antinuclear antibodies	Speckled morphology Title 1:80	Speckled morphology Title 1:160	Speckled morphology Title 1:80
Myositis specific autoantibodies (anti-synthetase, anti-Mi-2, anti- SRP)	Negative	Negative	Negative
Muscle MRI	Edema in shoulder and pelvic girdle and bilaterally in thighs and legs	Edema in shoulder and pelvic girdle and bilaterally in thighs and anterior legs	Edema in pelvic girdle and bilaterally in thighs and legs
EMG	Myopathic pattern	Myopathic pattern	Myopathic pattern
Muscle biopsy	Perimysial and perivascular infiltrates, predominantly macrophages	Interfascicular and perivascular infiltrates, predominantly T cells	Perimysial and perivascular infiltrates, predominantly macrophages

Table 2
Creatine kinase (CK) and myoglobin (Mb) serum levels in three patients with DM/PM.

	Patient A		Patient B		Patient C	
	CK (U/L)	Mb (μg/L)	CK (U/L)	Mb (μg/L)	CPK (U/L)	Mb (μg/L)
Admission values	2637	1089	4080	844	3041	734
Before starting TPE	1637	939	2880	645	2443	568
At the end of a TPE series	94	63	35	97	24	22
1 year after TPE was stopped	62	35	126	71	75	44

CK normal range: 20–180 U/L.
Myoglobin normal range: 12–70 μg/L.
TPE: Therapeutic plasma-exchange.

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