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Hypercapnic coma due to diaphragmatic involvement in a patient with dermatomyositis

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ABSTRACT

We report an acute respiratory distress by diaphragmatic involvement due to dermatomyositis. A fifty year-old patient with typical dermatomyositis presented an acute respiratory insufficiency with hypercapnic coma due to diaphragmatic muscle involvement. Respiratory state required mechanical ventilation initially and improved secondarily gradually upon corticosteroids and intravenous immunoglobulins. Only few cases of acute respiratory distress in dermatomyositis due to respiratory muscle involvement are reported in literature.

Introduction

Dermatomyositis (DM) is a rare inflammatory idiopathic myopathy of dysimmune origin. It associates muscular weakness and characteristic skin manifestations (1). Moreover dermatomyositis can be associated with connective tissue disease or malignancy (2). Corticosteroids are the main treatment but refractory polymyositis (PM) or dermatomyositis can require polyvalent human intravenous immunoglobulins or immunosuppressive agents (3, 4). An important cause of mortality or morbidity in inflammatory myopathies are respiratory complications (5-8). Nevertheless ventilatory insufficiency is rare in this cases. We report a case of acute respiratory distress with hypercapnic coma by diaphragmatic involvement requiring mechanical ventilation in a patient with dermatomyositis.

Case report

A 50-year-old woman was admitted to our internal medicine unit for an asthenia evolving for 4 months attributed to an hypothyroidism but unresponsive to hormonal substitutive therapy and an increase of hepatic enzymes. This important asthenia had led the patient to stop working. Upon admission, the typical lesions of dermatomyositis (Gottron's papules, liliaceous eyelids oedemas, periungueal erythema, subungual splinter hemorrhages) were evident, as well as a major general deterioration in her condition with a progressive weight loss of 10 kg over several months. Clinical examination showed muscle weakness – proximal in the upper limbs and proximal and distal in the lower limbs – associated with dysphagia. An interesting sign was observed – passing from the decubitus to the sitting position, there was a delay of the head on the trunk, as in the hypotonia of new-born babies who cannot raise their heads. We have called this sign the "new-born baby sign".

Biological assessment revealed an increase of hepatic enzymes, creatine phosphokinase serum level at 1848 IU (normal range: 30-135 IU), and lactate dehydrogenase at 2940 IU (normal range: 200-600 IU). The rest of the biological assessment found hypergammaglobulinemia at 18.7 g/L, a erythrocyte sedimentation rate of 20 mm, and normal renal function. The leucocyte count was 14,000/mm^3 with a lymphopenia of 840/mm^3. Tests of thyroid function found a subclinical hypothyroidism. The titre of antinuclear antibodies was 1/1280 without specificity, in particular without anti-Jo1. Rheumatoid factors were positive with a Waaler Rose of 1/128, a positive latex and IgM of 283 IU/ml by Elisa (normal < 20 IU/ml). The antineutrophil cytoplasmic, anti-Mi2, anti-LKM and anti-smooth muscles antibodies were negative. Chest radiography found an elevation of the left diaphragm. Electromyography showed myogenous syndrome and the muscular biopsy found inflammatory and necrotizing interstitial muscular involvement.

The patient fulfilled the diagnostic criteria for dermatomyositis (1). After treatment for 3 days with prednisolone 1 mg/kg per day, acute respiratory insufficiency appeared with coma (hypercapnia of 120 mm Hg, pH of 7.08) requiring transfer to the intensive care unit after an emergency intubation. The respiratory state was in relation with the diaphragmatic involvement of the DM without interstitial involvement on computed tomographic (CT) scan. Corticosteroid boluses were administered (240 mg per day intravenously for 3 days, then continuously at 1 mg/kg per day). However, in the absence of improvement, treatment with intravenous immunoglobulins (IVIG) (0.4 g/kg per day for 5 days) was begun. A very slow

favorable evolution was observed, allowing the weaning of assisted ventilation only at the second month. The patient presented the frequent complications of reanimation with infections and moderate decubitus ulcers, but her pulmonary state required corticoid maintenance therapy.

Once this difficult period had been passed, high-dose steroid treatment was maintained in association with monthly IVIG. This allowed a gradual weaning of corticoid therapy down to 15 mg per day at one year. Screening for a neoplasm by chest and abdomen CT scan, tumor markers and echography found a pelvic tumor. Surgical excision found a cyst dermoïde of the right ovary.

Recently a total weaning of steroid was obtained, but 1 month later the patient presented again with effort dyspnea, an increase of creatine phosphokinase serum levels from normal to 1283 UI, and the blood gas analysis found a PCO_2 of 52 mm Hg and a PO_2 of 55 mm Hg requiring the reintroduction of steroids at 1 mg/kg per day associated with IVIG treatment. The diaphragmatic involvement could be objectified by a magnetic resonance imaging of the diaphragm showing a very weak inspiration/expiration ratio of 1.08 (Fig 1). Pulmonary function tests showed significant restrictive respiratory insufficiency. The introduction of steroids and IVIG allowed a normalisation of the creatine phosphokinase serum level, a disappearance of dyspnea, and a gradual but incomplete improvement in the restrictive insufficiency at one month.

Discussion

Pulmonary involvement during polymyositis (PM) or DM can condition the prognosis. Interstitial lung disease is the most frequent form of pulmonary involvement, which can be expressed as infraclinic to severe dyspnea such as Hamman-Rich syndrome (5-8). The second cause of respiratory distress during PM and DM is aspiration pneumonia (5). Respiratory muscle involvement during PM or DM is said to be infrequent. It has been seldom evaluated in a systematic way, but frank respiratory insufficiency had been reported in 4 to 8% of DM and PM in various series (5). Respiratory muscle weakness generally becomes apparent in severe disease and is frequently associated with pharyngeal dysfunction. The ventilatory appears then progressively as anxiety or dyspnea and sometimes precipitous insufficiency as in our case (5). It is characterised by expiratory and inspiratory dysfunction. Hypercapnia did not appear until the respiratory muscle strength fell below 40% of normal (9). Measurement of the maximum respiratory pressures would seem to be important in monitoring corticosteroid response of patients with PM/DM, providing an objective measurement in addition to creatine phosphokinase, muscle testing and biological inflammatory parameters (10). Chronic respiratory failure due to respiratory muscle weakness has also been described in PM/DM but is more frequent in other muscle diseases such as the muscular dystrophies (8, 11). However, ventilatory insufficiency rarely results in an acute respiratory distress. Only some cases are reported in the literature and had been always associated with polymyositis (11-15). To our knowledge, this is the first report of respiratory muscle involvement with acute respiratory distress in a dermatomyositis.

In our case, an electrophysiologic study of the phrenic nerve might have been performed to eliminate the possibility of phrenic nerve involvement, but the severity of the muscle involvement, the correlation of the creatine phosphokinase serum level with the respiratory state at the moment of the relapse and on treatment, the well known diaphragmatic muscle involvement in PM/DM and the absence of phrenic nerve involvement reported in the literature in DM or PM did not in our eyes support the hypothesis of phrenic nerve involvement in this particular case.

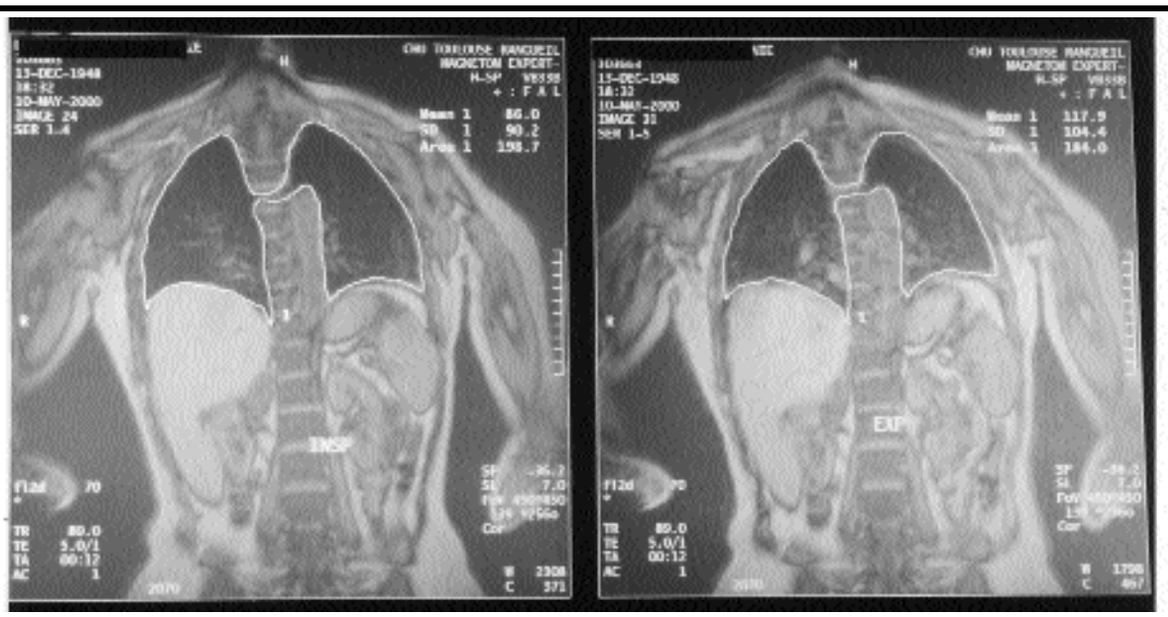

Fig. 1. Diaphragmatic MRI shows a very weak inspiration/expiration ratio of 1.08. The position of the diaphragm is almost the same during inspiration and expiration.

The use of IVIG is effective in refractory PM and DM (3,4) and could be an alternative treatment during mechanical ventilated DM in which infectious complications are frequent and may require a decrease or withdrawal of corticoids. Our observation illustrates the usefulness of IVIG in the course of DM with severe respiratory muscle involvement leading to acute respiratory insufficiency that required mechanical ventilation.

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