Juvenile dermatomyositis (JDM) and severe pulmonary involvement: case report

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ABSTRACT

Juvenile dermatomyositis (JDM) is an inflammatory, non-pustular disorder of skeletal muscles and skin. Other organs, such as the lungs, can be involved. Pulmonary complications are associated with high morbimortality rates and can develop in the acute phase of the disease. Due to its rarity, diagnosis difficulty and severity pulmonary involvement, we report the case of a 3-year-old girl with JDM and pulmonary involvement and evolution to death.

Keywords: juvenile dermatomyositis, diffuse alveolar injury.

INTRODUCTION

Juvenile dermatomyositis (JDM) is a non-suppurative inflammatory disorder of the skeletal muscles and skin. Other organs and systems, such as the lungs, gastrointestinal tract, subcutaneous tissue, eyes, and heart, can be involved.[1-6] Pulmonary complications are associated with high morbimortality rates and, usually, develop in the acute phase of the disease in up to 10% of the patients.[1,3,4]

Pulmonary involvement includes interstitial disease (pneumonitis or fibrosis), aspiration pneumonia, restrictive lung disease, ventilatory failure secondary to muscular weakness, infectious pneumonia, and drug-induced pneumonitis.[2,5]

Interstitial lung disease (ILD) seems to be the most common complication in adult patients with dermatomyositis (DM), affecting approximately 50% of the cases.[2,5-7] The main symptoms include coughing and dyspnea, but it can be asymptomatic in 5 to 50% of the cases of DM in the first years of the disease.[2,6,7]

Additional tests for the diagnosis of lung involvement include chest X-ray, multislice chest computerized tomography (MSCT), and pulmonary function tests (PFTs).[1,2]

There are few reports of patients with JDM and lung involvement. Most of them include severe cases, or cases refractory to treatment, which evolve to death.[1,3,4,6]

Due to its rarity, diagnosis difficulty and severity pulmonary involvement, we report the case of a 3-year-old girl with JDM and lung involvement and evolution to death.

CASE REPORT

Girl, 3 years and 9 months old, white, with a history of weight loss, progressive weakness, arthritis in the knees, skin rash with photosensitivity, and cutaneous ulcers in the tip of her fingers and ears for nine months. She had no fever and was hospitalized for pneumonia one month before this hospital admission.

On physical examination, her condition was regular, with decreased weight, mucous membranes were pale, she

Received on 12/15/2008. Approved on 02/22/2009. We declares no conflict os interest.
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had bilateral basilar crackles on lung examination, and hepatosplenomegaly; heart examination was normal. She also presented diffuse alopecia, vasculitis in the sole of her feet and nail folds, Gottron's sign, ulcers in the tip of her fingers and ears, Raynaud's phenomenon in the extremities, and bilateral arthritis of the knees, besides muscle weakness.

Laboratory tests revealed hemoglobin 10.7 g/dL, leukocytosis (12,900/uL), normal differential and platelet counts, erythrocyte sedimentation rate (ESR) 33 mm, aspartate transaminase (AST) 76 U/L (normal up to 32), alanine transaminase (ALT) 30 U/L (normal up to 31), lactic dehydrogenase (LDH) 550 U/L (normal up to 264), creatine kinase (CK) 31 U/L (26-140), aldolase 9.8 U/L (1.2-8.8), normal protein electrophoresis, negative antinuclear (ANA) and anticardiolipin antibodies, and normal complement and urinalysis. The myelogram and echocardiogram were also normal.

The chest X-ray showed a condensation on the right and bibasilar infiltrate. Muscular biopsy revealed perifascicular atrophy compatible with JDM. PFTs were not performed.

Fifteen days later, the patient was hospitalized again in another institution for a lung problem and, at that time, she was treated for three days with pulse methylprednisolone (30 mg/kg/day) and antibiotics, showing improvement of the vasculitis and lung problem. She was then treated with 1 mg/kg/day of prednisone. In that hospitalization, laboratory tests showed leukocytosis and neutrophilia, increased inflammatory activity tests, AST 200 U/L, ALT 96 U/L, LDH 917 U/L, CK 63 U/L, aldolase 12.5 U/L, and normal protein electrophoresis and urinalysis.

The echocardiogram showed ejection fraction of 0.58, diffuse myocardial hypokinesis, and a small pericardial effusion; ultrasound of the abdomen showed hepatomegaly.

Chest X-ray showed congestion, bibasilar infiltrate, and an increase in the size of the heart. Tomography of the chest revealed congested hila, severe infiltrate in the lower lobes, and increase in the size of the heart, but no pleural effusion.

The patient was discharged on naproxen (16 mg/kg/day) and prednisone (1 mg/kg/day). Three weeks later, the patient developed respiratory distress, tachycardia, and crackles in the right lung base without fever or worsening of the skin lesions. She also presented severe Raynaud's phenomenon in the extremities. The patient was admitted to the intensive care unit for cardiac and respiratory failure.

She evolved with fever spikes, and acute respiratory failure and a reduction in oxygen saturation requiring orotracheal intubation. Pulse methylprednisolone (30 mg/kg/day) was prescribed, but her respiratory function deteriorated progressively and the patient died three days after hospital admission. The chest X-ray was suggestive of alveolo-interstitial involvement (Figure 1). Urine and blood cultures were negative (including *fungi*), gastric washing was negative for alcohol-acid resistant bacilli, and serologies for mononucleosis, human immunodeficiency virus (HIV), and herpes virus were negative. Serology for IgG and IgM cytomegalovirus (CMV) was positive; anti-Jo 1 negative.

Lung lesions with alveolar hemorrhage, hyperplasia type II pneumocytes, and hyaline membrane, characterizing diffuse alveolar damage, were observed on autopsy. Those lesions, associated with infectious pneumonia, lead to acute respiratory distress, cardiac ischemia, circulatory collapse, and death (Figure 2). Indications of lung infection with CMV were not observed.

DISCUSSION

JDM is a systemic autoimmune vasculopathy that affects, mainly, the skin and muscles, with a characteristic cutaneous rash and proximal myopathy, which can also affect other organs, such as the gastrointestinal tract and lungs, which are associated with uncertain prognosis.[1-6]

The patient presented here had diagnostic criteria for JDM:[8] symmetrical muscular weakness, cutaneous vasculitis, increased muscle enzymes, and muscle biopsy compatible with JDM.

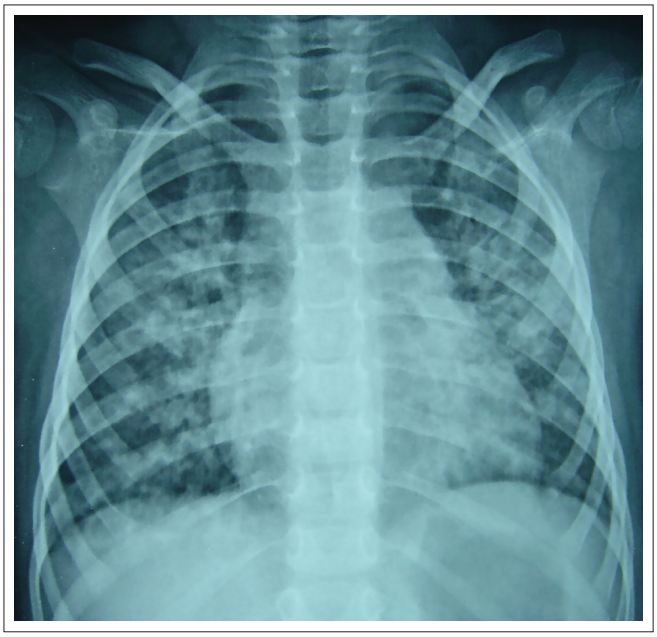

Figure 1. Chest X-ray with diffuse interstitial infiltrate and increased heart area.

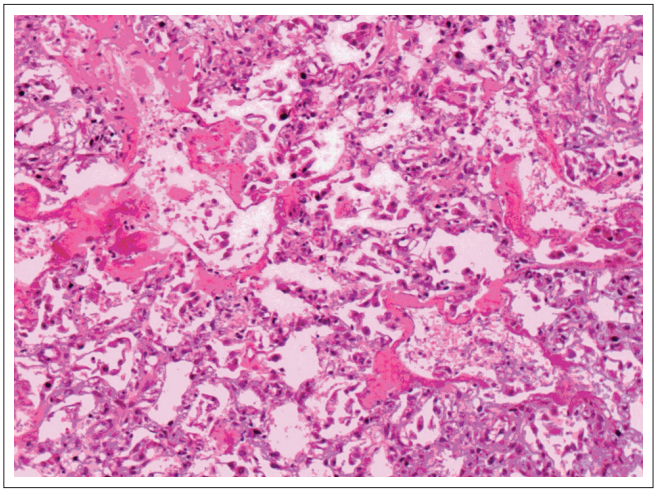

Figure 2. Diffuse alveolar involvement. Hyaline membrane, proteinous matter, inflammatory interstitial infiltrate, and pneumocyte hyperplasia can be seen.

There are few reports of lung involvement in children with JDM,[1,3,4,6] which is associated with a worse prognosis and can manifest even before the development of cutaneous and musculoskeletal signs and symptoms.[1,3,4]

In a study by Trapani *et al.*[1] with 12 patients with JDM, 50% of the patients had asymptomatic lung disease detected by PFTs.

The presentation of ILD can be acute or chronic, associated with coughing, dyspnea, and lung infiltrate. It can be asymptomatic with altered PFTs and normal imaging exams.[2] In the study performed by Kang et *al.*[9] diffuse alveolar damage and interstitial pneumonia in the lung biopsy were associated with a worse evolution of the ILD in adults with DM and polymyositis. In the patient presented here, the lung evaluation during autopsy showed diffuse alveolar damage and pneumonia, which could have contributed for the acute deterioration of the respiratory function.

The association of ILD with the Jo1 autoantibody, characterizing the antisynthetase syndrome, has been reported.[1,2,3,5,9] This syndrome is rare in children, and it is characterized by non-erosive arthritis, Raynaud's phenomenon, fever, and interstitial lung disease, with severe dyspnea and pulmonary fibrosis. It is associated with a poor prognosis, with a 70% survival in five years.[1] The patient presented here had non-erosive arthritis, Raynaud's phenomenon, and interstitial lung involvement. Anti-Jo1 was negative, but it can be negative in up to 69% of the patients with DM and ILD.[3,10]

Despite the positive CMV serology, *postmortem* tissue staining did not show indications of lung infection with CMV. The autopsy results were suggestive of infectious pneumonia, but the etiological agent was not identified.

The use of cyclosporine and cyclophosphamide, with improvement of lung function in adults and children with ILD, has been reported.[6,11] However, although it was indicated in the patient in question, pulse cyclophosphamide was not used due to the fulminant course of her disease.

The presence of ILD in patients with JDM is associated with a poor prognosis. Chest X-ray and high-resolution tomography, pulmonary function tests, and anti-Jo1 should be done in patients with JDM for an early diagnosis of lung involvement and treatment.

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