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Case Report

A Case of Juvenile Dermatomyositis Presenting with Respiratory Distress

Gürkan Gürbüz[1], Gulden DİNİZ[2] and Aycan Unalp[1]*

[1]Health Sciences University, Dr. Behcet Uz Childrens Training and Research Hospital Izmir, Turkey

[2]Neuromuscular Disease Center, Tepecik Training and Research Hospital, İzmir

***Corresponding Author**: Aycan Unalp, Professor of Pediatric Neurology, SBU, Dr. Behcet Uz Child Disease and Pediatric Surgery Training and Research Hospital, Department of Pediatric Neurology, Izmir, Turkey.

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Abstract

Juvenile dermatomyositis is an autoimmune inflammatory myopathy that presents with a heliotropic rash over the eyelids, cutaneous eruptions on the extensor sides of the limbs, and proximal muscle weakness. Diagnosis is usually made based on clinical findings. Muscle biopsy can be helpful in cases that do not fulfill the diagnostic criteria.

A 6-year-old girl with muscle weakness who had been hospitalized in the pediatric intensive care unit for over 3 weeks developed newly arising respiratory distress. A physical examination and laboratory and muscle biopsy data indicated that she had juvenile dermatomyositis. Both the respiratory distress and muscle weakness responded well to immunosuppressive therapy. We report the case because such a presentation in uncommon in childhood.

Keywords: *Juvenile Dermatomyositis; Respiratory Distress; Myopathy*

Introduction

Juvenile dermatomyositis (JDM) is an autoinflammatory autoimmune myopathy, and is the most common inflammatory myopathy of childhood. The annual incidence is 2-3/1,000,000 children and the average age at onset is 7 years [1]. Dermatological and muscle manifestations are typical, as are a distinct heliotropic rash on the eyelids, an eczematous rash (Gottron papules) on the extensor surfaces of the extremities, and proximal muscle weakness. JDM is usually diagnosed clinically, accompanied by microscopic examination and laboratory and serological testing. Muscle biopsy is also diagnostic. The differential diagnoses include other myopathies, hereditary and acquired neuropathies, and late-onset metabolic disorders. We here present a case of JDM in a 6-year-old child with primary respiratory distress, which is rare among JDM cases.

Case Report

A previously healthy 6-year-old girl became increasingly fatigued over a 3-month period when climbing stairs, developed a cough 2 weeks in duration, and experienced increasing respiratory distress; she was referred to us from a local hospital.

On first examination, her general status was poor. The Glasgow Coma Scale score was 15 and her body weight 15 kg (fifth percentile). Her height was 109 cm (tenth percentile); body temperature 36.6°C; heart rate 127 beats/min; respiratory rate 40 breaths/min; and oxygen saturation 88% (despite delivery of 5L oxygen/min through a mask). No crackles or rhonchi were evident on auscultation of the thorax; the breath sounds were bilaterally decreased in the basal zones; and intercostal and subcostal retractions were observed. On neurological examination, the deep tendon reflexes were normoactive in the upper extremities but areflexia was apparent in the lower extremities. Gower's sign was present. The muscle strengths of both lower extremities were 3/5 (on a scale of 0 - 5) proximally and 4/5 distally; the upper extremity scores were 4/5 proximally and 5/5 distally. Both the direct and indirect pupillary light reflexes were reactive. An eczematous rash was evident on the proximal interphalangeal and metacarpophalangeal joints of both hands, and a heliotropic rash on the eyelids. Other evaluations were normal. Although oxygen was delivered at 5 L/min through a mask, the respiratory distress problems persisted and the oxygen saturation was < 90%. Non-invasive mechanical ventilation was commenced.

Her creatine kinase (CK), alanine aminotransferase (ALT), and aspartate aminotransferase (AST) levels were all elevated (2,200 IU, 172 mg/dL, and 153 mg/dL, respectively). Both the erythrocyte sedimentation rate (70 mm/h) and C-reactive protein level (2.35 mg/dL) were also elevated. Arterial blood gas analysis revealed respiratory acidosis: pH 7.25, pCO_2 65%, pO_2 70%, and HCO_3 35%. No other pathology was evident on laboratory testing. A chest X-ray revealed infiltrates in the right paracardiac region compatible with interstitial pneumonia.

The absence of deep tendon reflexes in the lower limbs suggested ascending paralysis; we thus considered Guillain-Barre syndrome and JDM during differential diagnosis. She underwent craniospinal magnetic resonance imaging (MRI), which eliminated the possibility of a lesion in the intramedullar space and a demyelinating pathology. Lumbar puncture was performed; the cerebrospinal fluid (CSF) biochemical data were normal (glucose 69 mg/dL, Cl 105 mmol/L, protein 20 mg/dL). Microscopic examination of the CSF revealed no erythrocytes or leucocytes.

The purple swollen appearance of the metacarpophalangeal and proximal interphalangeal joints of the fingers, the elevated creatine kinase (CK) level (see below), and myopathic changes evident on electromyography were suggestive of JDM; immunoglobulin was administered intravenously (IVIG) at 1 g/kg/day for 2 days. Cefotaxime was commenced to treat the pneumonia. The respiratory distress increased during follow-up, and a repeat X-ray revealed effusion in the right lower lung that was confirmed by thoracic ultrasonography. The effusion was 5 mm in width and was considered parapneumonic in nature; the antibiotherapy was switched to cefepime and clindamycin. High-resolution computed tomography (HRCT) was performed. Focal consolidations combined with air bronchograms were evident in the superior, posterior, and basal segments of the lower lobe of the right lung and the posterior and basal segments of the lower lobe of the left lung. Mild tubular bronchiectasis was apparent in the lower lobes of both lungs. Minor sequelae were detected in the anterior segment of the right upper lobe. A comprehensive viral panel test and the TORCH panel test were negative. Serological tests for S*almonella, Brucella* and *Mycoplasma* were negative. The anti-ganglioside antibody panel test used to evaluate Guillain-Barre syndrome-spectrum disorders was negative. Basal metabolic tests evaluating late-onset metabolic disease status were also normal. Serum CK status was evaluated in the context of suspected myopathy and was higher than the first reading, at 3,300 IU/mL. This elevation persisted, and myopathic changes were detected on electromyography (EMG) after the respiratory distress had eased. The antinuclear antibody (ANA) test was positive at 1:100, and exhibited a nucleolar pattern. The extractable nuclear antibody (ENA) panel test was negative, including the anti-Jo test.

The right quadratus femoris muscle was biopsied to confirm the diagnosis just before steroid treatment commenced. The biopsy revealed inflammatory myopathy with perivascular infiltration. No typical feature of dermatomyositis (such as perifascicular atrophy) was apparent in the biopsy specimen, perhaps attributable to the heterogeneous nature of the disease. The biopsy findings were consistent with JDM, and intravenous pulsed methylprednisolone (30 mg/kg/dose, never exceeding 1 g/dose) was commenced. After 7 days, the pulse therapy was discontinued and oral steroids (1 mg/kg/day) commenced. On day 15 of treatment, the CK level was 620 IU, and fell to 310 IU on day 20. At this time, the patient could walk and deep tendon reflexes were present in all extremities. In addition to the steroid, we commenced weekly methotrexate. Chest X-ray revealed that the infiltrations and effusions had regressed. We consider that the pneumonia had developed secondary to weakness of the respiratory muscles. As pulmonary fibrosis with interstitial involvement may develop secondary to dermatomyositis, we scheduled close follow-up. The patient was discharged on weekly subcutaneous methotrexate therapy, which is the most effective treatment for JDM.

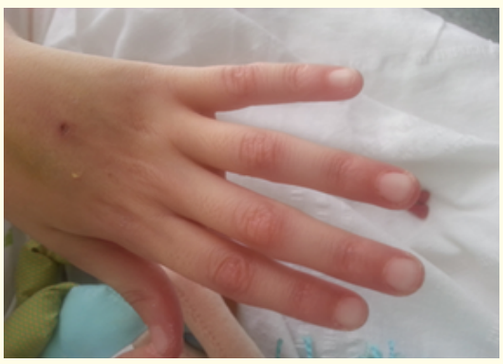

Figure 1: *Gottron Papules on PIP and MCP joints.*

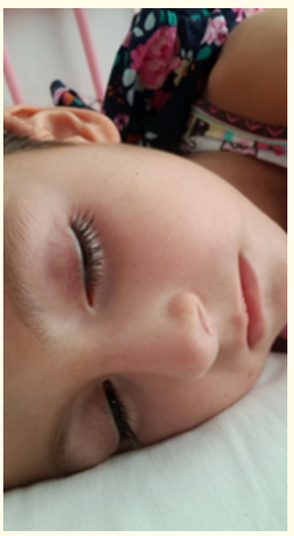

Figure 2: *Heliotrope Rash on eyelids.*

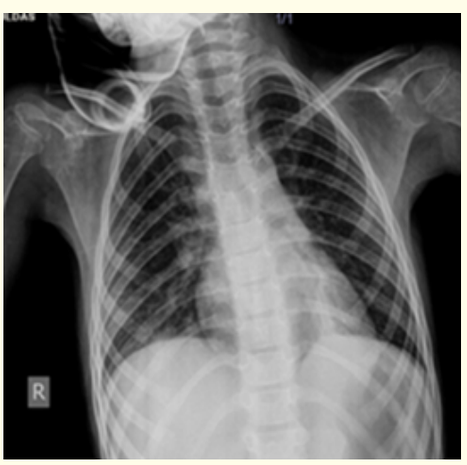

Figure 3: *Interstitial pneumonia in X-Ray.*

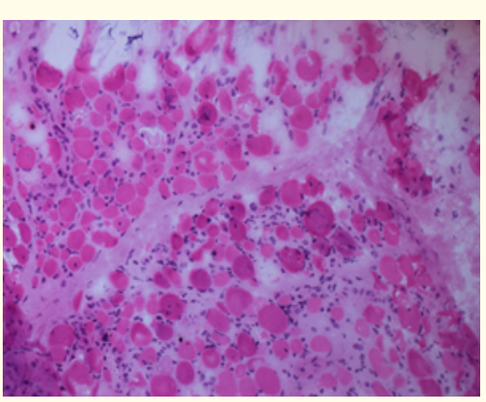

Figure 4: *Differences in the size and shape of myofibers, regenerated fiber, inflammation.*

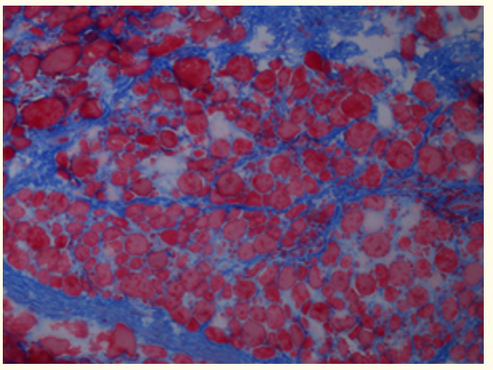

Figure 5: *Increased interstitial tissue, Trichrome Stain.*

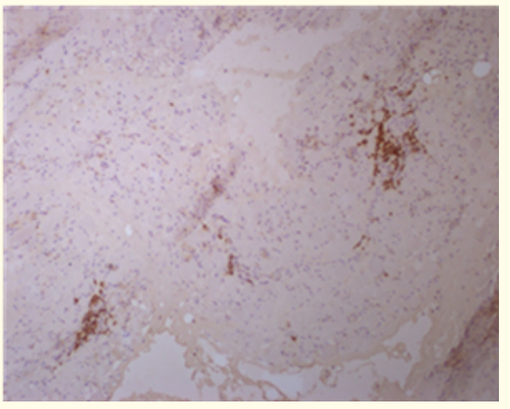

Figure 6: *Microscopic appearance of inflammatory mostly CD3 T cells.*

Discussion

JDM is diagnosed using clinical, laboratory, and microscopic methods. Bohem and Peter previously described the diagnostic criteria. The classical criteria (of which three or four must be met) include dermatological findings, myopathy evident on EMG, typical dermatomyositis features in a muscle biopsy specimen, elevated levels of muscle enzymes (CK, lactate dehydrogenase, AST, and aldolase), and symmetrical muscle weakness [2,3]. Our patient fulfilled four of these criteria.

Today, diagnosis is assisted by neuroimaging strategies, including muscle ultrasonography (US) and MRI. US reveals muscle edema and swelling, and MRI muscle edema and any form of heterogenous involvement [4]. US and MRI data are not listed in the diagnostic criteria but are nonetheless very useful.

Muscle biopsy is helpful when a diagnosis is vague, or when only a suspicion is raised. As most patients exhibit heterogeneous muscle involvement, typical histological features of disease (perivascular lymphocytic infiltration, scattered myeloid cell infiltration, endothelial/small cell abnormalities, muscle spindle regeneration, and necrosis) may be absent [5]. Tuen., et al. used MRI to place guidewires in affected muscle groups to facilitate excisional biopsy in cases where the pathology is patchy or dispersed. We biopsied the muscle group that was most affected, as revealed by EMG. We were unable to directly target a particular muscle, for technical reasons. The disease is heterogeneous, and we did not note the perifascicular atrophy typical of JDM. The absence of this feature does not preclude a diagnosis of JDM.

Our patient was ANA-positive (1:100) and exhibited a nucleolar pattern. All of ANA, anti- Jo1, anti-p155/140, and anti-signal recognition particle antibodies may be detected in JDM patients. Anti-Jo1 antibody is associated specifically with interstitial lung disease. The ANA test is positive in 80% of patients, but does not aid in the differential diagnosis of JDM from scleroderma or polymyositis.

The extent of lung involvement is 5 - 45% in adults and may develop secondary to reflux, aspiration pneumonia, pleural effusion, spontaneous pneumothorax, interstitial fibrosis, or respiratory muscle dysfunction [6]. In our present case, the pulmonary infection developed secondary to respiratory muscle dysfunction, and the pleural effusion secondary to infection. The pleural effusion induced respiratory distress. Pleural fluid sampling was not possible because the effusion was < 10 mm in width. It is unclear whether the effusion developed secondary to infection or was rather a feature of a rheumatic condition.

Many reports suggest that JDM patients may develop chronic lung disease in adulthood. Respiratory distress caused by primary or secondary lung involvement is rare in children at the time of first admission to treat JDM [5-8].

Myopathic changes evident on EMG are diagnostic of JDM. When the patient was initially admitted, her general status was poor. She suffered from respiratory distress and required non-invasive mechanical ventilation. Thus, EMG could not be performed and IVIG therapy (2 g/kg) was commenced. Treatment options for JDM include IVIG, methotrexate, rituximab, cyclophosphamide, and corticosteroids (oral or intravenous). Corticosteroids are not recommended as long-term treatments because of side-effects. Corticosteroids should be combined with second-line therapy. After 7-day intravenous pulsed methylprednisolone (30 mg/kg/dose, never exceeding 1 g/dose), the patient was switched to oral prednisolone (2 mg/kg/day) combined with subcutaneous methotrexate (10 mg/m2 weekly) [7,9]. We also administered IVIG at 1 mg/kg/day on 2 days. Initially, we avoided steroid therapy because of the pulmonary infection.

Prior to the advent of immunosuppressive therapy, the prognosis of JDM was poor (mortality was about 33%); prognosis improved with the introduction of corticosteroid and immunosuppressive agents (mortality has fallen to < 1%). The extent of disease activity is reflected by the clinical assessment and the serum levels of muscle enzymes. The presence of muscular fibrosis and atrophy render it difficult to evaluate muscle strength. The levels of serum muscle enzymes are good indicators of disease activity. Recent studies have shown that measurements of IL-6, IL-8, and TNF-alpha are useful during follow-up [10,11]. Clinically, the levels of serum muscle enzymes decreased after corticosteroid therapy in our patient.

Conclusion

Although dermatomyositis is the most common inflammatory myopathy of childhood, we thought it worthwhile to present a rare case featuring respiratory distress. We suggest that JDM should be kept in mind during the differential diagnosis of flaccid paralysis accompanied by respiratory distress.

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