Juvenile Dermatomyositis; Clinical and Laboratory Assays

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Abstract

Objective: Objective: Juvenile dermatomyositis (JDMS) involves children rarely. Our purpose of this study is to detect clinical and laboratory aspects of this disorder and its treatment in Khorasan Province.

Material & Methods: A 10-years retrospective study performed on medical records of all patients with JDMS in a medical center in Mashhad. Data was analyzed with statistical measures including SPSS and Excel.

Findings: 18 patients had DJMS during 10 years. There was a female to male predominance (1.25/1). The mean age of patients was 12.1 years. Muscular weakness was the most common presenting symptom. Skin involvement including heliotrope rash and Gottron papules were detected in most patients. Elevated muscular enzymes were an important finding in many cases. EMG, and muscle biopsy in those cases performed, showed abnormal results. Seventeen (94.5%) of patients responded to oral prednisolon therapy.

Conclusion: According to clinical manifestations and laboratory findings, in particular muscular enzyme assays, a timely diagnosis and treatment with oral prednisolon can improve the disease dramatically and reduce morbidity and mortality rates as well.

Key Words: Juvenile dermatomyositis, Children, EMG, Muscle enzymes, Skin

Introduction

Juvenile dermatomyositis (JDMS) is a rare disease. The incidence of JDMS is 2 to 4 per million children [¹]. It is a chronic inflammatory disorder which presents with proximal muscle weakness and a characteristic skin rash. It is seen in children aged 2 to 15 years [²,³]. Generally, it occurs before 18 years of life [⁴]. Laboratory findings consist of elevation of muscle enzymes such as CK and LDH, myopathic changes in EMG, and inflammatory changes in muscle biopsy.
In one study from India, all patients had proximal muscle weakness at presentation [5]. Another survey from Hungary revealed that most cases had skin manifestations consisting of facial erythema and Gottron papules [6]. Also, another study from Thailand demonstrated raised levels of muscle enzymes and myositis changes in all JDMS patients [7].

JDMS is an important collagen vascular disease. A timely diagnosis and prompt treatment can reduce the morbidity and mortality rates significantly. There was no extensive study done in Khorasan Province, and in practice we found some differences between the characteristics of our cases and those of patients in some other countries including average age of onset and the most common initial complaint. So, we decided to review the clinical and laboratory findings as well as methods of treatment in our patients. This is the first extensive retrospective study of JDMS in Khorasan Province.

**Material & Methods**

This is a 10-year retrospective study performed on 18 cases of JDMS admitted to Ghaem and Imam Reza hospitals in Mashhad. Data including age, gender, clinical and laboratory findings, and treatment were arranged in special sheets. We included in our study only those records that had definite diagnosis based on clinical and laboratory findings. Cases with probable diagnosis of Juvenile dermatomyositis were omitted from study. Information was analyzed with SPSS and Excel.

**Findings**

There were 18 patients, 10 females and 8 males (F/M:1.25/1). Table 1 shows the incidence of JDMS in various age groups. Mean age of the patients at presentation was 12.1 years. On physical exam, all patients suffered from muscle weakness. Eleven patients (61.1%) had weakness in proximal muscle of lower limbs, 6 patients had pharyngeal muscle weakness and one case showed flexure neck muscle weakness. The most common presenting symptom was muscle weakness. Table 2 shows the causes and frequency of presenting complaint in our patients.

Seventeen patients (94.4%) had skin rash, 9 cases (50%) with heliotrope rash and 8 patients (44.4%) with Gottron papules in metacarpophalangeal and phalangeal-interphalangeal. Arthralgia and arthritis were demonstrated in 16 patients (88.9%). Two cases (11.1%) had calcinosis. Seventeen percent of cases had axillary lymphadenopathy. No hepatosplenomegaly was detected in our patients. Chest x-ray was normal in almost all of our cases. Only one patient had positive guaiac test.

Laboratory findings showed mild leukocytosis in 3 cases, elevated ESR in 66.6% of cases and abnormal CRP in 27.5% of patients. Rheumatic

<table>
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<tr>
<th>Age group (years)</th>
<th>Absolute frequency (%)</th>
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<tbody>
<tr>
<td>4 - 5</td>
<td>0</td>
</tr>
<tr>
<td>6 - 7</td>
<td>3 (17%)</td>
</tr>
<tr>
<td>8 - 9</td>
<td>0</td>
</tr>
<tr>
<td>10 - 11</td>
<td>4 (22%)</td>
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<tr>
<td>12 - 13</td>
<td>4 (22%)</td>
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<tr>
<td>14 - 15</td>
<td>7 (39%)</td>
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<table>
<thead>
<tr>
<th>Symptom</th>
<th>Absolute frequency (%)</th>
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<tbody>
<tr>
<td>Muscle weakness</td>
<td>14 (78%)</td>
</tr>
<tr>
<td>Muscle pain</td>
<td>5 (28%)</td>
</tr>
<tr>
<td>Arthralgia+ Arthritis</td>
<td>4 (22%)</td>
</tr>
<tr>
<td>Skin rash including heliotrope rash</td>
<td>3 (17%)</td>
</tr>
<tr>
<td>Extrimities edema</td>
<td>2 (11%)</td>
</tr>
<tr>
<td>Periorbital edema</td>
<td>1 (6%)</td>
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Factor (RF) and anti-nuclear antibody (ANA) were abnormal in 12.5% of cases. Table 3 shows other laboratory results.

EMG was performed in 11 patients and all of them had abnormal results. Muscle biopsy in 7 patients was performed which was compatible with myositis. Skin biopsy was performed in 5 patients, revealed vasculitis changes.

All of our patients received high dose oral prednisolon, but one patient (5.5%) did not have improvement and received hydroxyquin. Neither cytotoxic agents nor IVIG was begun for our cases. Generally, after 15-20 days of beginning of prednisolon, clinical symptoms improved and a significant reduction in muscle enzymes was detected. Nine patients, who are at remission, receive minimum dose of prednisolon. One case took one year prednisolon and then the drug was withheld successfully. One case, that had calcinosis with disturbing consequences, had improvement by surgery. Other cases did not return to clinic after discharging from hospital.

**Discussion**

According to our results, JDMS is more common in females than males (1.25/1). Mean age of patients at diagnosis was 12.1 years old. In one study form North India mean age of patients was 8.7 years ± 3.3 [8]. In another study from France male to female ratio was 0.75 and mean age of patients was 7.7 years old [9]. Also, another report from Spain shows that female to male ratio is 2.1 and 7 years old is mean age of diagnosis of cases [10].

Presenting complaint is muscle weakness says our results. Similar finding is reported from France, Thailand and Spain [9,7,10]. In Indian study, all cases had muscle weakness or pain at admission [5]. Survies from Hungary, America and Saudi Arabia have shown that all of patients suffered from symmetrical proximal muscle weakness [6,11,12].

Fifty percent of our patients revealed heliotrope rash and 44% Gottron papules. It was like the report from Saudia Arabia in which study heliotrope rash was detected in 52% of cases and Gottron papules in 60% [12]. Also, Hungarian report demonstrated that 11 of 12 cases had facial erythema and Gottron papules [6]. According to thai study, 85% of cases showed heliotrope sign and 28% of cases Gottron papules [7].

Eighty percent of our patients suffered from arthritis and arthralgia. Saudia Arabia survey revealed that 64% of their patients had arthritis [12]. Hungarian study, demonstrated arthralgia in 58% of JDMS [6]. Thirty five percent arthritis in patients was reported from American study in JDMS [11].

Calcinosis was detected in 11% of our cases. It was near to Sao Paulo study (11.42%) [3]. Other reports from Thailand, America and India revealed the occurrence of calcinosis 56%, 23% and 16% respectively [7,11,5].

Alimentary involvement was 5% in ours and 17.4% in Sao Paulo report [13]. There was an increased muscle enzyme in our cases. 14 of 15 patients (93%) had raised LDH, 10 of 16 cases (62.5%) raised CK, 6 of 7 patients (58.5%) increased Aldolase and 11 out of 14 (78.5%) cases raised AST. It is similar to results from Sao Paulo study [13]. American study revealed CK rise in 60%, LDH in 73, AST in 69% and Aldolase in 31% of cases [14]. Spanish survey showed that CK and LDH rise in 89% of cases, Aldolase and AST in 77.5% and 66% respectively [10].

In our study, ESR in 66.6% and ANA in 12.5% were abnormal. ESR in 80% of cases and ANA in 43% of patients were elevated in regard to study from America [14]. Auto antibodies were undetectable in Spanish review [10].

EMG was performed in 11 of our cases with abnormal results in all of them. Myositis was the finding in all 7 cases who had muscle biopsy. Seventy nine percent of cases revealed abnormal

<table>
<thead>
<tr>
<th>Test</th>
<th>Absolute frequency (%)</th>
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<tr>
<td>Increased AST</td>
<td>11 (61%)</td>
</tr>
<tr>
<td>Rised CK</td>
<td>10 (56%)</td>
</tr>
<tr>
<td>Rised Aldolase</td>
<td>6 (33%)</td>
</tr>
<tr>
<td>Increased LDH</td>
<td>14 (78%)</td>
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EMG changes and 11 of 16 cases with muscle biopsy also showed myositis or atrophic changes, according to survey from USA [14].

In Spain report all of cases who had EMG, demonstrated myopathic changes. Also inflammatory myopathy changes showed in that cases who performed muscle biopsy [10]. Another study in America revealed that 19% of patients had normal EMG and 20% of cases who had muscle biopsy, showed normal result as well [11].

After treatment with high dose oral prednisolon, 94.5% had complete improvement and one patient, who did not have improvement, received hydroxyquin. Similar results reported from Thailand, where oral prednisolon (1-2 mg/kg/day) was associated with complete recovery [7]. Successful therapy with oral prednisolon also was reported from Spain and India [8,10]. Also 88% of cases in Sao Paulo review had good response to oral prednisolon [13].

**Conclusion**

In conclusion, attention to muscle weakness and skin rash in children under 16 years old and rapid diagnosis and prompt treatment with oral prednisolon after confirmation of diagnosis, can reduce morbidity of this disorder obviously.

**References**