

Juvenile Dermatomyositis in Arab Children: A Multi-Center Retrospective Analysis of Disease Features and Outcome

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Abstract: Objective

To describe disease characteristics and outcome of Arab children with juvenile dermatomyositis (JDM)

Methods

We retrospectively reviewed children with JDM seen between 1990 and 2014 in 5 rheumatology clinics from 3 Arab countries. All included patients fulfilled Bohan and Peter criteria for JDM, diagnosed before 14 years of age and were Arab ethnicity. Data were collected at the last follow up visit and comprised of age of presentation, follow up duration, clinical and laboratory features, treatment as well as the long-term outcomes including accrual disease damage and death related to JDM.

Results

A total of 89 JDM patients (55 girls) from Saudi Arabia (65.2%), Jordan (18%) and Oman (16.9%) were included. The mean age at onset was 6 years (+ 3), the mean age at diagnosis was 6.6 years (+ 3), and with mean follow up duration of 5 years (+ 4.4). Fifty patients had active follow up, 27 patients transferred to adult rheumatology service and 12 patients had lost follow up. Forty patients had polycyclic disease course, 34 had monocyclic course while 15 had continuous progressive course. Seventy-four patients had organ involvement, arthritis (48%), upper airway and dysphagia (14%), gastrointestinal (13%) and lung involvement (10%). Seven patients admitted to intensive care unit (ICU), 4 of them required mechanical ventilation. Dysphagia, upper airway and lung involvement were all statistically significant associated with ICU admission. All patients received corticosteroids; while methotrexate was the most frequently used immunosuppressive drugs (86%) and rituximab was used in 8 patients. Additionally 31 patients received IVIG. Twelve patients received pamidronate mainly for calcinosis. Most of the patients achieved complete clinical response but 16 ended with permanent skin changes and 12 had residual muscle weakness. Twenty-seven patients developed calcinosis, 15 had osteoporosis, and 3 had lipodystrophy while 37 patients had growth failure. One patient developed mucinous cystic ovarian tumor. There were two deaths due to infection during the follow up period.

Conclusion

Arab patients with JDM have similar characteristics to previously described cohorts. However, compared to our previous experience, a steadily improvement in outcome particularly calcinosis which probably related to better therapeutic approach.

Key words: Juvenile dermatomyositis, Arab, Treatment, Outcome