

Spectrum of cases in juvenile Dermatomyositis (JDM)

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Abstract

Objective

1. To raise awareness about JDM
2. To explore different presentations in JDM
3. How to approach a case of JDM

Contents

- Cases (Clinical characteristic and diagnosis of different Cases of Libyan children with JDM follow up at rheumatology department since 2011 - 2019 on Tripoli children hospital / LIBYA)
- Diagnosis

Juvenile Dermatomyositis is a rare, potentially life-threatening systemic autoimmune vasculopathy of the skin and muscle. The most common pediatric inflammatory myopathy (85%) causing symmetric proximal weakness and a characteristic skin rash (heliotrope, Gottron papules, ulcer, scar and calcinosis). It affects about 3 in 1 million children each year, F: M ratio 2: 1 bimodal age distribution (ages 5-9 year and 10-14 year)

In 1975, Bohan and peter proposed a diagnostic criterion for JDM

1.Symmetrical weakness of the proximal muscles 2. Characteristic cutaneous changes: heliotrope, per orbital edema, Gottron papules 3. Elevation of serum muscle enzymes 4. EMG show characteristics of myopathy 5. Muscle biopsy: inflammation and or vasculitis

Outlines of management the main treatment of JDM

Corticosteroids (1-2 mg/kg per day) initially used to induce remission over 4-6 weeks with a slow taper. Weaning may take 2 years

- Many of patient fail to respond adequately to corticosteroids and require additional immunosuppressive medication as IVIG, methotrexate, cyclosporine, rituximab)

International PRINTO group

To facilitate the follow-up of children with JDM, recently, **the international PRINTO group - (Pediatric Rheumatology International Trials Organization)**- reviewed the measures indicated to evaluate the treatment of the disease and recommended the following: 1) global disease assessment by the physician 2) evaluation of muscle strength 3) global assessment of disease activity 4) global well-being assessment by the parents 5) functional capacity 6) health-related quality of life. All those have discriminative capacity, internal consistency, good validity and are easy to use

Article Information

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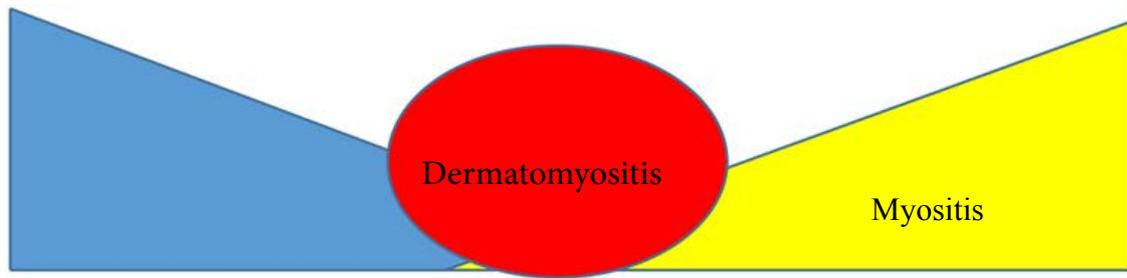
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Dermatitis

Conclusion

- JDM presented with different spectrum of cases Dermatitis, myositis and both Dermatomyositis
- Overlap syndrome: JDM overlap with many CT disease as SLE scleroderma, JIA

- Early diagnosis and aggressive treatment directed at achieving rapid and complete control of muscle inflammation is highly successful in minimizing the long term sequel of JDM, including calcinosis

Keywords

CT disease: connective tissue disease, SLE: systemic lupus erythematosus, JIA: juvenile idiopathic arthritis