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內文：

Introduction

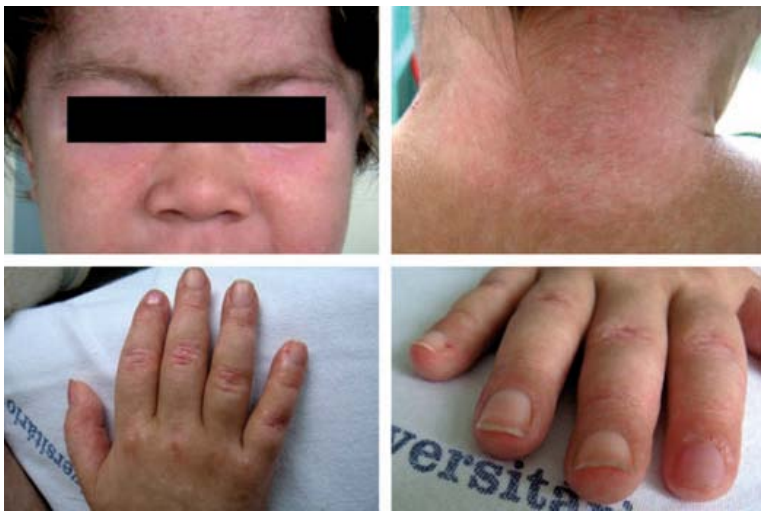
Juvenile dermatomyositis (JDM) is a rare **inflammatory disease of skeletal muscle** with characteristic **skin manifestations**. It is considered the most common form of myopathy in patients between 2 and 18 y/o.

Case report

A 4 y/o girl presented to her pediatric dentist for evaluation of gingival tenderness and bleeding and white patched in her tongue. Two weeks later, she developed a **facial eruption**, myalgias, muscle weakness, and **difficulty in rising from a sitting position**, which prompted a referral to the hospital.

Physical examination

(1) Heliotrope, photosensitive facial rash
(2) Hyperkeratotic erythematous patches on elbow, legs, and upper back
(3) Papules over the interphalangeal joints of the fingers (Gottron papules)
(4) Ragged cuticles with dilated nail-fold telangiectases

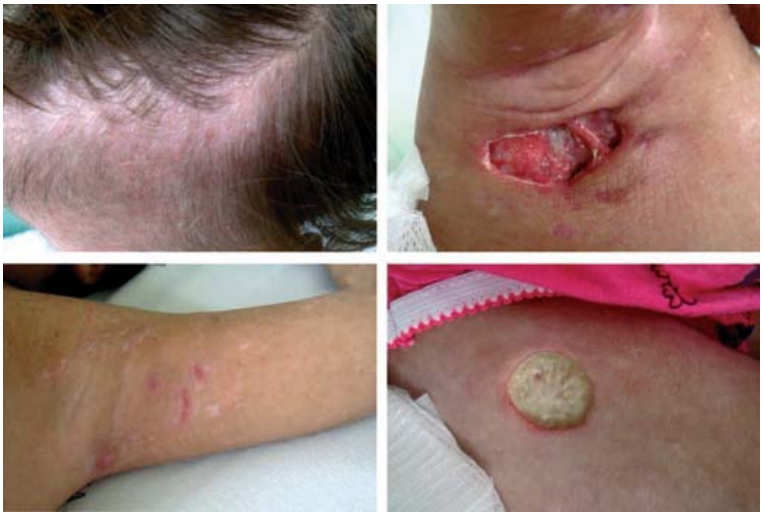


(5) Diffuse alopecia

(6) Genital edema

(7) Subcutaneous calcifications throughout her body indicated to surgery (E.g. extensive calcinosis in axilla)

(8) Necrotic ulceration on the thigh



(9) Complicated **erysipelas** on the leg (confirmed by bacteriological examination)

Oral examination

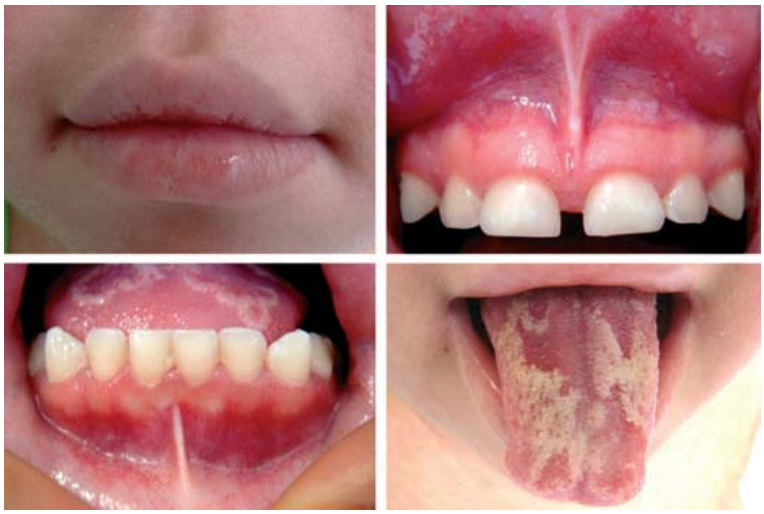
(1) Lip edema and prominent **erythema**

(2) Multiple dilated telangiectases along the lower gingiva

(3) Intermittent episodes of depapillated and erosive patches with tongue that resemble migratory glossitis

(4) Halitosis

(5) **Dysphagia** with consequent **weight loss** and **asthenia**



Differential diagnosis

(1) Vesiculo-bullous disease
(2) LE
(3) Lichen planus
(4) Lichenoid reaction to atenolol

No blistering was associated with the ulceration and the skin lesions were not typical of any of the above condition.

Lab data

(1) Complete blood count (CBC): normal
(2) Routine chemistries: normal
(3) Muscle enzymes levels indicated a high inflammatory activity
   a. **CPK level** of 795 U/L (normal ≦ 185 U/L)
   b. **Aldolase level** of 70 U/L (normal ≦ 7.6 U/L)
(4) Electoneuromyography (EMG): a typical pattern of myogenic injury

Muscle biopsy: consistent with JDM

(1) Muscle fiber: atrophy, necrosis
(2) Lymphocytic infiltrate

Treatment

(1) 1st to 3rd day: methylprednisolone pulse therapy (30 mg/kg per day)
(2) 4rd day to 3nd month: prednisone (2 mg/kg per day) +
chloroquine diphosphate (5 mg/kg per day)
3rd month: muscular and cutaneous findings, as well as the gingival disease had substantially improved
(3) 4th month - : prednisone (1 mg/kg per day)
(4) 1-year f/u: prednisone (0.5 mg/kg per day)
continue to improve, regaining muscle strength of the neck, abdominal flexors, and proximal upper and lower extremities

Comment

JDM is a multisystem disease characterized by **acute and chronic inflammation of the skeletal muscle and skin**.

Prevalence: 1 – 3.2 cases per million in children

Diagnosis: presence of characteristic cutaneous changes and three of the following four criteria:

(1) Symmetric weakness of the proximal musculature
(2) Elevation of the serum level of skeletal muscle enzymes
(3) Electromyographic demonstrations (EMG)
(4) Positive muscle biopsy

This case report illustrates a classical form of JDM. The patient fulfilled all of diagnostic criteria for confirming the clinical, histological, EGM, and biochemical diagnosis. However, an outstanding feature is the unusual represented the **first signs** of the condition. Although a biopsy of our patient's oral lesions was not carried out, they were believed to be oral JDM for a number of reasons:

(1) The **unique appearance and distribution** of the lesions
(2) Their time course **shortly to the first cutaneous and muscular findings**
(3) The **response** of oral lesions **to treatment**

Differential diagnosis

Cutaneous manifestations and muscle weakness:

(1) Acute allergic contact dermatitis
(2) Photodermatitis
(3) Seborrhoic dermatitis
(4) Atopic dermatitis
(5) Polymorphic light eruption
(6) Systemic LE
(7) Subacute cutaneous LE
(8) Lichen planus
(9) Psoriasis
(10) Orbital cellulites
(11) Cutaneous T cell lymphoma

Oral manifestations:

(1) Vesiculo-bullous disease
(2) LE
(3) Lichen planus
(4) Lichenoid reaction to atenolol

Only one case of JDM with tongue involvement hase been published, which traced a patient similar to ours, where the child had intermittent episodes of depapillated and erosive tongue.

Capillary abnormalities in the gingiva have been described in five patients with JDM and the authors felt that they were analogous to the **periungual telangiectases** that are seen in the nail beds of patients with this disease, suggesting that oral symptoms are important diagnostic markers. These oral lesions may be as an **initial manifestation of the disease,** which leads us to propose that an early identification is essential for an immediate treatment.

Pediatric dentists may play a primary role in the diagnosis of JDM. The importance of investigations in this area is determined by the fact that the disorder may have **acute evolution and may finish with fatal outcome**, or in other cases, with **chronic, protracted March that leads to serious invalidity**, and in the final stage needs special care.

Conclusion

Knowledge od the pediatric dentist about the oral lesions, as well as its associations with systemic alterations, may be essential in the **early identification of conditions such as JDM**, which contribute in the **diagnosis and installation of appropriated treatment**, consequently favoring the prognosis.

題號	題目
1	Which of the following is not a clinical feature of perioral dermatitis? (A) Papules (B) Papulopustules (C) Pruritus (D) Edema
答案 (D)	出處：Brad W. Neville, Douglas D. Damm, Carl M. Allen, Jerry E. Bouquot. *Oral and Maxillofacial Pathology*. 3rd ed. St. Louis, MO: Sounders; 2009: 352.
題號	題目
2	Which of the following is not an antibiotic used to treat perioral dermatitis? (A) Tetracycline (B) Amoxicilline (C) Erythromycin (D) Metronidazole
答案 (B)	出處：Brad W. Neville, Douglas D. Damm, Carl M. Allen, Jerry E. Bouquot. *Oral and Maxillofacial Pathology*. 3rd ed. St. Louis, MO: Sounders; 2009: 352.