DOI: 10.1111/j.1365-263X.2010.01074.x

INTERNATIONAL JOURNAL OF PAEDIATRIC DENTISTRY

Oral manifestations as important symptoms for juvenile dermatomyositis early diagnosis: a case report

LETÍCIA MACHADO GONÇALVES[1], JOSÉ RIBAMAR SABINO BEZERRA-JÚNIOR[1], MANUEL ANTONIO GORDÓN-NÚÑEZ[2], SILVANA AMADO LIBÉRIO[3], ADRIANA DE FÁTIMA VASCONCELOS PEREIRA[4] & MARIA CARMEN FONTOURA NOGUEIRA DA CRUZ[4]

[1]Department of Dentistry, School of Dentistry, Federal University of Maranhão, São Luís, MA, Brazil, [2]Department of Oral Pathology, School of Dentistry, Federal University of Rio Grande do Norte, Natal, RN, Brazil, [3]Department of Dentistry I, School of Dentistry, Federal University of Maranhão, São Luís, MA, Brazil, and [4]Department of Dentistry II, School of Dentistry, Federal University of Maranhão, São Luís, MA, Brazil

International Journal of Paediatric Dentistry 2011; 21: 77–80

Background. Juvenile dermatomyositis (JDM) is an idiopathic inflammatory myopathy of childhood and adolescence, characterized by symmetrical weakness of proximal muscles and classical cutaneous features. Literature reports rarely describe or focus on oral lesions that are associated with this disease.

Case report. This case describes a 4-year-old girl in whom the oral lesions were the initial manifestations of JDM. Physical examination revealed characteristic skin manifestations, proximal muscle weakness, extensive calcinosis, necrotic ulceration, complicated erysipelas, and diffuse alopecia. The diagnosis was established based on the clinical, histological, electroneuromyography, and biochemical findings.

Conclusion. Recognition of gingival telangiectases as an important diagnostic marker of JDM leads us to suggest that identifying oral manifestations, which may be carried out by a paediatric dentist, contributes in establishing an early diagnosis and an immediate treatment of this condition.

Introduction

Juvenile dermatomyositis (JDM) is a rare inflammatory disease of skeletal muscle with characteristic skin manifestations[1–3]. It is considered the most common form of myopathy in patients between 2 and 18 years old[3–6]. Several reports have focused on the oral manifestations of the adult form of dermatomyositis (DM)[7–10], but the recent literature has rarely described the oral findings of JDM[4,8]. The juvenile form is frequently associated with gingival telangiectases and this has been proposed as an underappreciated diagnostic finding[8,10]. Alterations in tongue have been rarely documented[8,9].

Correspondence to:
D. L. M. Gonçalves, Department of Dentistry II, Federal University of Maranhão, Avenida dos Portugueses, s/n, Bairro Bacanga, São Luís, MA 65085-580, Brazil.
E-mail: lets.mg@gmail.com

Case report

A 4-year-old girl previously in good health presented to her paediatric dentist for evaluation of gingival tenderness and bleeding and white patches 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.

Results of physical examination were remarkable for a heliotrope, photosensitive facial rash (Fig. 1a) and hyperkeratotic erythematous patches on elbow, legs, and upper back (Fig. 1b). Other signs included papules over the interphalangeal joints of the fingers (i.e., Gottron papules; Fig. 1c), ragged cuticles with dilated nail-fold telangiectases (Fig. 1d), diffuse alopecia (Fig. 2a), and genital oedema. Subcutaneous calcifications, such as extensive calcinosis in axilla, throughout her body were noted (Fig. 2b), and indicated to surgery

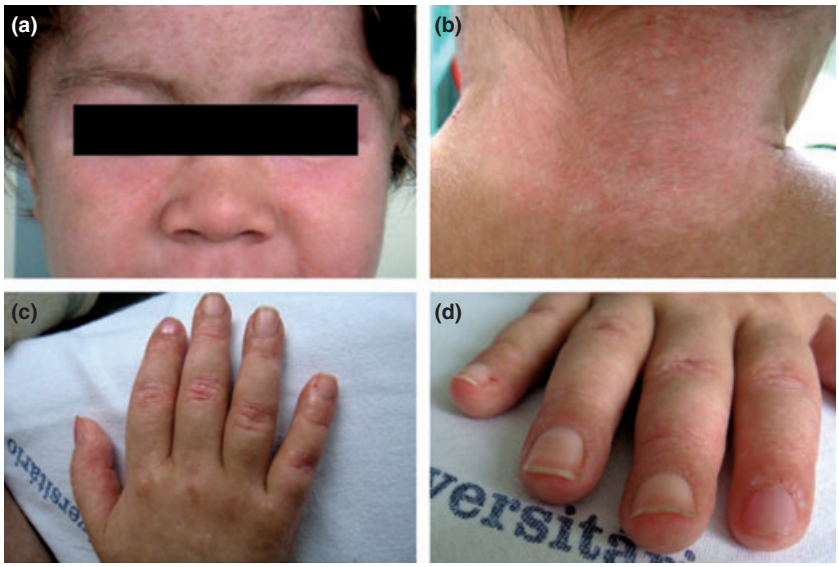

Fig. 1. Classical cutaneous features detected at the physical examination: (a) heliotrope and facial rash, (b) hyperkeratotic erythematous patches on upper back, (c) Gottron's papule overlying interphalangeal joints of the fingers, (d) dilated periungual telangiectases and ragged cuticles, with little points of bleeding.

(Fig. 2c); necrotic ulceration on the thigh (Fig. 2d) and complicated erysipelas on the leg were confirmed by bacteriological examination.

The results of oral examination revealed lips oedema (Fig. 3a) and prominent erythema, consisting of multiple dilated telangiectases along the lower gingiva (Fig 3b,c). The child also had intermittent episodes of depapillated and erosive patches with thick margins over the dorsum of the tongue that clinically resembled migratory glossitis (Fig. 3d). In addition, she presented halitosis and dysphagia, with consequent weight loss and asthenia.

The most likely causes of the oral lesions were a vesiculo–bullous disease, lupus erythematosus, lichen planus, or a lichenoid reaction to atenolol. However, no blistering was associated with the ulceration for this patient and the skin lesions were not typical of any of the above conditions.

Complete blood count and routine chemistries were within normal ranges. However, the muscle enzymes levels indicated a high inflammatory activity with CPK level of 795 U/L (normal ≤185) and aldolase level of 70 U/L (normal ≤7.6 U/L). Electroneuromyography (EMG) demonstrated a typical pattern of myogenic injury. Muscle biopsy specimen showed muscle fibre atrophy, necrosis, and lymphocytic infiltrate histologically consistent with JDM.

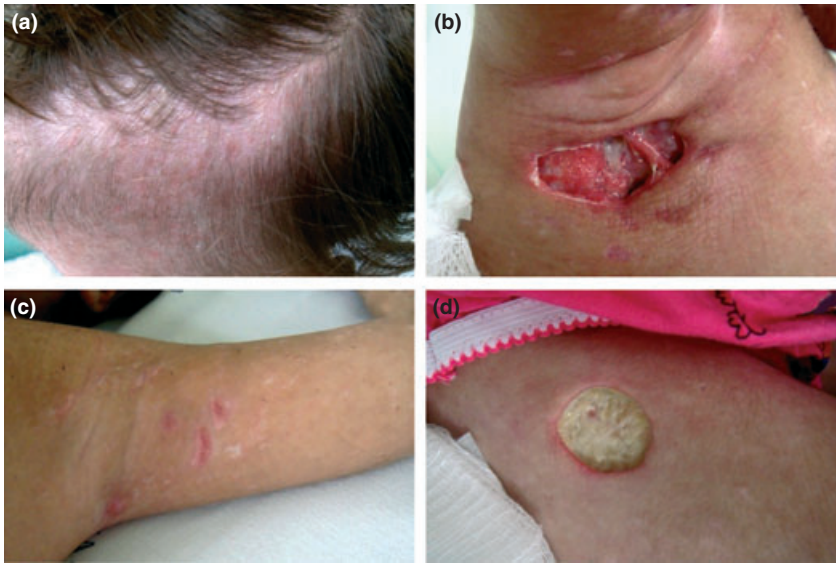

Fig. 2. Other signs observed: (a) diffuse alopecia affecting a big part of the scalp, (b) subcutaneous calcifications on arms, (c) axillary region after surgical intervention to remove extensive calcinosis, (d) necrotic ulceration on the right thigh.

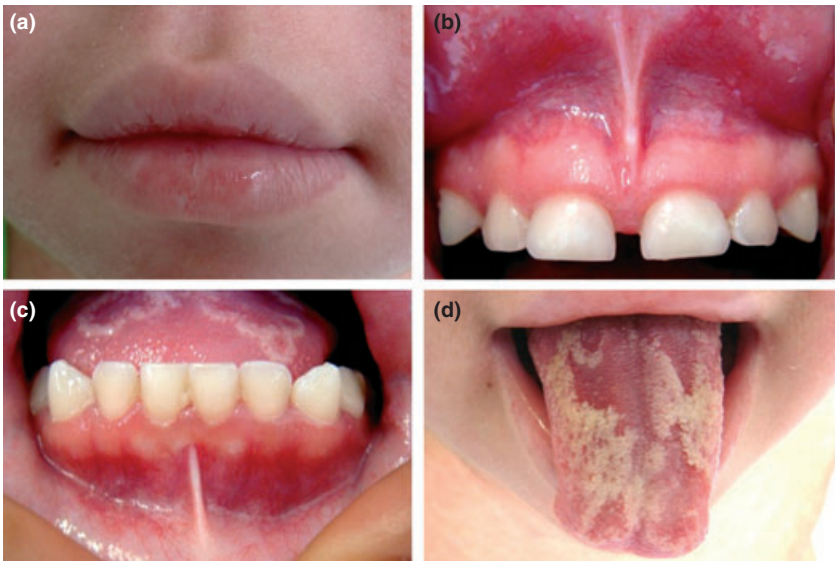

Fig. 3. Signs observed after oral examination: (a) lips oedema; (b) and (c) dilated telangiectases on the lower gingiva and on the neighbouring mucous membranes in both dentitions; (d) depapillated and erosive patches over the dorsum of the tongue, resembled migratory glossitis.

The diagnosis of JDM was confirmed and treatment was started with methylprednisolone pulse therapy (30 mg/kg per day, for three consecutive days) followed by maintenance of prednisone (2 mg/kg per day) combined with chloroquine diphosphate (5 mg/kg per day). Two months later, the muscular and cutaneous findings, as well as the gingival disease, had substantially improved. The prednisone dose was tapered to 1 mg/kg per day over 3 months. At 1-year follow-up, while on a tapering prednisone regiment (0.5 mg/kg per day), she continued to improve, regaining muscle strength of the neck, abdominal flexors. and proximal upper and lower extremities.

Comment

Juvenile dermatomyositis is a multisystem disease characterized by acute and chronic inflammation of the skeletal muscle and skin. The disorder is rare, with a prevalence of 1–3.2 cases per million in children[1,2]. Besides the presence of characteristic cutaneous changes, diagnosis of JDM requires three of the following four criteria: symmetric weakness of the proximal musculature, elevation of the serum level of skeletal muscle enzymes, electromyographic demonstrations (EMG), and a positive muscle biopsy[6].

This case report illustrates a classical form of JDM. The patient fulfilled all of diagnostic criteria for confirming the clinical, histological, EMG, and biochemical diagnosis[6]. However, an outstanding feature is the unusual presence of oral manifestations, which 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 DM for a number of reasons: the unique appearance and distribution of the lesions; their time course shortly to the first cutaneous and muscular findings; and the response of oral lesions to treatment.

There is a wide differential diagnosis for the clinical symptoms of cutaneous manifestations and muscle weakness in JDM patients. Differential diagnosis of early skin manifestations of JDM include acute allergic contact dermatitis, photodermatitis, polymorphic light eruption, systemic LE, subacute cutaneous LE, seborrhoic dermatitis, lichen planus, psoriasis, orbital cellulites, cutaneous T cell lymphoma, and atopic dermatitis[10]. In addition, differential diagnosis of oral manifestations includes vesiculous–bullous diseases, lupus erythematosus, lichen planus, or lichenoid reaction. The absence of these disorders in the present case strongly suggested that these lesions were related to JDM.

The earliest reports of oral involvement in JDM described erythema and telangiectases at the gingival margins[4,8], whereas lesions of tongue are less frequently reported[8]. To the

best of our knowledge, only one case of JDM with tongue involvement has been published[8], 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[8] 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. Nevertheless, these oral lesions may be significant not only merely in identifying subsets of JDM, but also as an initial manifestation of the disease, which leads us to propose that an early identification, both by the paediatric dentist and/or the dermatologist, is essential for an immediate treatment.

Paediatric dentists may play a primary role in the diagnosis of JDM. The importance of investigations in this area is determined by the fact that, regardless of the restricted number of JDM patients with oral involvement, 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. Serious prognosis and high mortality rate of the disease ranges from 3% in JDM in the USA[4]. As exemplified, oral symptoms may precede the evolution of skin lesions and myositis, and in these cases, paediatric dentist should refer patients as quickly as possible to receive specialized medical care.

It should be emphasized that oral involvement is underappreciated in JDM. The frequency of these lesions, as well as their natural history and correlation with muscle damage needs to be further defined. Despite the fact that oral manifestations are not often recognized, the case related is a rare entity. In conclusion, it is believed that knowledge od the paediatric dentist about 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 favouring the prognosis.

What this paper adds

- Recording a very rare inflammatory myopathy of childhood and adolescence.
- Recognition of oral manifestations as an important finding for JDM diagnosis.
- Suggesting that gingival telangiectases may represent the first signs of JDM.

Why this paper is important for paediatric dentists

- Gingival telangiectases may represent the first signs of JDM and an early identification, which may be carried out by the paediatric dentist, is essential for the diagnosis and immediate treatment.
- Adding a new clinical experience for JDM cases is important, as these cases are extremely rare.

Acknowledgements

This study was partly supported by Fapema, São Luís, MA, Brazil.

Conflict of interest

All authors have declared no conflict of interest.

References

1 Callen JP. Collagen vascular diseases. *J Am Dermatol* 2004; **51**: 427–439.
2 Chiu SK, Yang YH, Wang LC, Chiang BL. Ten-year experience of juvenile dermatomyositis: a retrospective study. *J Microbiol Immunol Infect* 2007; **40**: 68–73.
3 Sallum AM, Kiss MH, Sachetti S *et al.* Juvenile dermatomyositis: clinical, laboratorial, histological, therapeutical and evolutive parameters of 35 patients. *Arq Neuropsiquiatr* 2002; **60**: 889–899.
4 Pachman LM, Boskey AL. Clinical manifestations and pathogenesis of hydroxyapatite crystal deposition in juvenile dermatomyositis. *Curr Rheumatol Rep* 2006; **8**: 236–243.
5 Peloro TM, Fred Miller O III, Hahn TF, Newman ED. Juvenile dermatomyositis: a retrospective review of 30-year experience. *J Am Acad Dermatol* 2001; **45**: 28–34.
6 Bohan A, Peter JB. Polymyositis and dermatomyositis. *N Engl J Med* 1975; **292**: 344–347, 403–407.
7 Keil H. The manifestations in the skin and mucous membrane in dermatomyositis, with special reference to the differential diagnosis from systemic lupus erythematosus. *Ann Intern Med* 1942; **16**: 828–871.
8 Ghali FE, Stein LD, Fine J-D *et al.* Gingival telangiectases: an underappreciated physical sign of dermatomyositis. *Arch Dermatol* 1999; **135**: 1370–1374.
9 Healey CM, Tobin AM, Kirby B, Flint SR. Oral lesions as an initial manifestation of dermatomyositis with occult malignancy. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2006; **101**: 184–187.
10 Dourmishev LA, Dourmishev AL, Schwartz RA. Dermatomyositis: an association of gingival telangiectases and anti Jo-1 antibody in the adult. *Acta Dermatoven APA* 2007; **16**(2): 67–72.