

Oral signs in juvenile dermatomyositis

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Abstract:

Juvenile Dermatomyositis (JDM) is a severe inflammatory myopathy characterized by multisystemic vasculopathy during childhood. The general profile consists of symmetric proximal muscle weakness and the presence of characteristic rashes. This case describes an 11-year-old girl with hyperplastic gingival inflammation, telangiectasia and lichenoid retroalveolar striae detected by dental examination visit, when she also showed rash and papules in her hands. Histopathological diagnosis of the retroalveolar mucosa was indicative of unspecific chronic inflammation. Later when muscle symptoms and serum changes were present the JDM diagnosis was confirmed. The patient was successfully treated with systemic corticosteroids and regular professional dental cleaning. In order to get an early diagnosis and effective treatment for the patients, it is important that oral health care providers are aware of the oral manifestations related to systemic diseases, such as JDM.

Keywords: Dermatomyositis; Telangiectasis; Gingiva; Mouth Mucosa.

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INTRODUCTION

Juvenile dermatomyositis (JDM) is a systemic vasculopathy of unknown etiology, characterized by acute and chronic inflammation of the skeletal muscle and skin^{1,2}. Although rare, JDM is the most common subset of the juvenile idiopathic inflammatory myopathies (JIIMs), representing about 85% of the cases among children³. Advances in treatment with corticosteroids and immune suppressants have reduced the mortality rate of JDM to 1-5%, but the disease is still a cause of significant morbidity⁴.

Clinically, JDM is characterized by muscle weakness and typical skin involvement. It may also involve multiple other organs, including the gastrointestinal tract, heart, lungs, kidneys and eyes¹. The orofacial region can also be affected, but only few studies have focused on its changes in JDM⁵⁻⁷. However, oral lesions can be initial manifestations of the disease, and especially gingival telangiectasia has been proposed as an important diagnostic marker⁸. Here we present a case of JDM, where the oral mucosa changes together with rash in the hands were the first findings and indicative of diagnosis.

CASE REPORT

An 11-year-old girl was referred to the Oulu University Dental School due to the gingivitis which did not response to the traditional treatment. She had an erythematous generalized lesion in the attached and free gingiva of both upper and lower jaws (Fig. 1A). An accurate intraoral examination showed dilation of the gingival capillaries close to teeth crowns, and also bilateral erythema and non-detachable white lichenoid striae in the retroalveolar mucosa (Fig. 1B and D).

The patient had intact teeth without any dental restorative material. A biopsy from the retroalveolar gingiva was carried out. The tissue H&E staining showed unspecific pseudoepitheliomatous epithelial hyperplasia, mild chronic inflammation and was signed out as a "suspected as a lupus-like reaction" (Fig. 1C). The physical examination was remarkable for her hands, presenting papules and erythematous rash in the distal, proximal and metacarpophalangeal joints, compatible with Gottron's papules (Fig. 2A).

The medical history of the patient was non-significant. Because of the skin findings the patient was referred for a skin biopsy. The tissue H&E staining showed epithelial hyperplasia and mild chronic inflammatory infiltrate within the connective tissue (Fig. 2B). A frozen



Figure 1. JDM patient at the first appointment had generalized erythematous free gingiva with multiple telangiectasia (A); whitish lichenoid striae in the both retroalveolar mucosal areas, and telangiectasia in the posterior teeth gingiva, presenting also some spontaneous bleeding points (B and D). Biopsy specimen was taken from the area marked as a black circle in B; it shows slight epithelial hyperparakeratosis, pseudoepitheliomatous hyperplasia and a connective tissue with mild chronic inflammation and permeated small vessels (C, x100). Biopsy was signed out as "suspected lupus-like reaction".

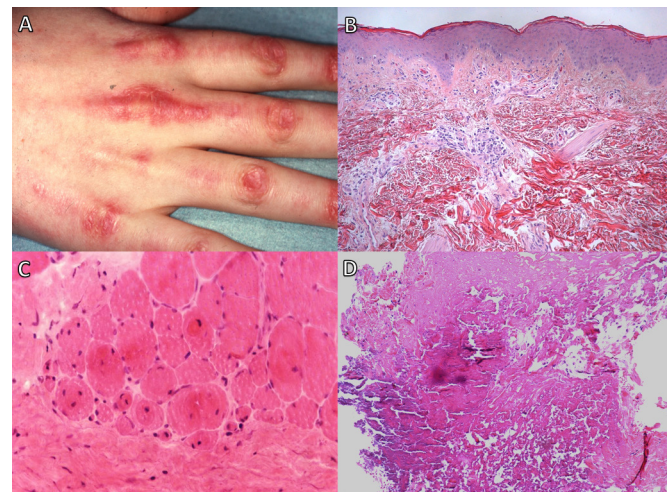


Figure 2. Characteristic Gottron's papules in the fingers joints (A). Histological section from one of the papules showing epithelial hyperplasia and mild chronic inflammatory infiltrate within the connective tissue (B, x100). Mild myositis and necrosis are shown in the section of muscle biopsy (C, x200). Calcified tissue removed from the lower jaw gingiva presented some inflammatory cells with areas of dystrophic calcification (D, x200).

tissue section was analyzed by immunofluorescence for Polyclonal Igs, IgG, IgM, IgA, C3 and fibrinogen to rule out autoimmune diseases, such as lupus erythematosus. The immunofluorescence findings were negative. A couple of months later the patient developed muscle symptoms. Serum analysis showed the presence of Jo-1 autoantibodies and a biopsy of muscle tissue revealed mild myositis and necrosis (Fig. 2C).

The diagnosis of JDM was confirmed based on the clinical, histological and serum findings. The treatment was based on systemic corticosteroids (prednisolone 50 mg daily) and oral cleaning by dental hygienist every two months. The dermatological changes became worse in the middle of the treatment and prednisolone doses were increased.

After the remission of the signs and symptoms the steroid was gradually stopped. After the treatment, the skin appearance improved and muscle power was within normal limits. Despite the main remission, enlarged and condensed tissues in the gingival regions of the teeth 43-44 and 33-36 developed, and histopathological analyses showed necrotic areas with dystrophic calcifications within connective tissue (Fig. 2D). After 23 years of follow-up the patient has remained with no recurrent disease.

DISCUSSION

JDM is the most common phenotype among the JIMs (onset before 16 years old), with an annual incidence from 1.9-4 cases per million children, depending on the population⁹⁻¹³. Several studies have shown a female predominance, in a ratio as high as 5:1 in a British survey, and an average onset of 7 years^{1,9,11,14-17}. The disease seems to have a bimodal behavior in girls with two peaks, before 6 and around 11 years old (when our patient was diagnosed), but a single peak around 10 years in boys^{11,18}.

Symmetric proximal and axial muscle weakness and the presence of characteristic rashes, i.e., Gottron's papules (erythematous plaques overlying the extensor joint surfaces) or heliotrope rash (a purplish or erythematous rash over the eyelids) are general characteristics of JDM. In addition, the patients often have other photosensitive rashes, including malar erythema, photosensitivity, linear extensor erythema, as well as other cutaneous findings^{19,20}.

Besides the presence of the pathognomonic rash, diagnosis of JDM requires three of four Bohan and Peter criteria: proximal muscle weakness, elevated serum levels of muscle enzymes, electromyographic changes of chronic inflammatory myositis, and histopathological changes of inflammatory myositis^{7,21}. Our patient presented the characteristic rash in her hands, muscle symptoms, serum changes and myositis confirmed by biopsy, which taken together led to a JDM diagnosis.

Interestingly, the patient was referred to the clinic reporting a "gingivitis" resistant to the traditional approaches. The first oral signs of disease were prominent erythema and telangiectasia along gingival margins and lichenoid lesion in retroalveolar mucosa. Later on,

a calcified tissue in the lower maxillary gingiva was biopsied. Prominent dilated capillaries along the facial anterior marginal gingiva has been reported previously as one of the diagnostic signs of JDM.

The gingival vasculopathy pattern associated with hyperplasia and gingival bleeding, distinct from periodontal disease and associated with cutaneous disease activity, suggests that gingiva is a possible target tissue for JDM⁶. Differing from the adult form, the pathogenesis of JDM can be based on vasculopathy rather than simply an inflammation of skin and muscle sites¹⁸.

The Table 1 summarizes our case and all previously published JDM cases presenting oral involvement. The most common oral mucosa findings were telangiectasia, erythematous lesions and dysphagia, which can be related to the capillary changes. JDM patients seem to share a unique gingival pattern characterized by gingival erythema, capillary dilation and bush-loop formation⁶.

Similar to the lesion presented in retroalveolar area in our patient, Ghali *et al.*⁵ described reticulated whitish patches on the buccal mucosa and tongue of a 7-year-old girl diagnosed with JDM. Another patient presented depapillated and erosive patches over the dorsum of the tongue⁷. Ulcers, whitish and erosive patches are changes also described in patients with the adult form of dermatomyositis^{28,29,30}. In the case presented here, the lesion regressed after systemic corticosteroid therapy, indicating close relation with the general disease. The relation between oral whitish/lichenoid lesions and dermatomyositis (juvenile and adult form) needs further investigation.

Calcinosis are not rare in JDM and can occur in 20-40% of the patients¹⁹. Cutaneous calcinosis is frequently located on the elbows, knees, and the acral and proximal parts of extremities, whereas calcification of oral soft tissues are uncommon. Only one case of oral soft tissue calcinosis related to JDM has been reported before: bilateral, superficial and painful subcutaneous protrusions, visualized in x-ray as symmetric calcifications in the mandible²⁶. In our case, the calcified gingival tissue was surgically excised and had no remission. Calcified focal points in facial bones should be investigated for systemic diseases such as JDM²⁶.

In the recent years, mortality due to JDM has declined considerably but morbidity related to the myopathy, calcinosis and long-term use of corticosteroids remains a major challenge⁹. Although the majority (50-60%) of patients with JDM experience a chronic illness course, approximately 24-40% of the patients have a

Table 1. Review of cases presenting juvenile dermatomyositis with oral involvement.

First author	Published year	Number of cases	Age (y)	Sex	Oral features	Concomitant disorders
Present case	-	1	11	F	Erythematous attached and free gingiva, telangiectasia bilateral white lichenoid striae in the retroalveolar mucosa, gingival calcinosis	-
Carbonell ²²	2015	1	12	F	Dysphagia	-
Ferreira ²³	2012	1	10	F	Burning in the mouth; leucoplakia lesions on the tongue; pain when chewing and swallowing	Chronic hyperplastic candidiasis
Gonçalves ⁷	2011	1	4	F	Gingival tenderness, multiple dilated telangiectasia and bleeding; depapillated, erosive, and white patches with thick margins over the dorsum of the tongue that clinically resembled migratory glossitis; lips oedema; halitosis; dysphagia	-
Rider ⁸	2009	1	16	F	Dysphagia; bleeding gums; marked dilation of the capillaries of the attached gingiva	-
Zedan ²⁴	2008	1	3.5	M	Dysphagia, swollen lips, and drool	-
Holmes ²⁵	2008	1	15	M	Lymphoepithelial sialadenitis	Sjogren's syndrome
Sarifakioglu ²⁶	2003	1	19	F	Painful subcutaneous protrusions at the mandible angle level (bilateral soft tissue calcifications in x-ray)	-
Ghali ⁵	2001	5	8	M	Gingival tenderness, bleeding, discomfort, erythema, telangiectasia, and swelling; asymptomatic depapillated atrophic patches with thick white margins over the dorsum of the tongue	-
			11	F	Gingival erythema and telangiectasia	
			9	F	Gingival erythema and telangiectasia	
			5	F	Oral discomfort; gingival erythema (composed of individual dilated telangiectasia) and inflammation; whitish and reticulated patches on buccal mucosa and tongue	
			10	F	Dysphagia; oral discomfort; gingival severe erythema composed of multiple telangiectasia	
Mitchell ²⁷	2001	1	7	F	Dysphagia	-

Note: JDM cases were only included when presenting oral involvement and complete data from all patients. (M: Male; F: Female).

monocyclic course, recovering with appropriate therapy within a 2-year period¹⁹. Since our patient did not show recurrence after 16 years from the treatment, we consider that she has fully recovered. Oral health care providers should be aware of the JDM signs in oral cavity, specially aiming to early diagnosis of this systemic disease. As in our case, the oral and skin findings followed by muscle and blood analysis led rapidly to the right diagnosis of JDM.

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