Juvenile dermatomyositis with gingival vasculopathy

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Presentation

A 6-year-old boy with juvenile dermatomyositis (JDM) positive for anti-TIF1 γ antibodies for 3 years on methotrexate (1 mg/kg) reported with recurrent heliotrope and Gottron's rashes, and muscle weakness refractory to corticosteroids (1 mg/kg) for 3 months. He progressively developed periorbital edema, oral ulcers, and skin ulcers over the trunk. On examination, Gottron's papules, periungual desquamation, ulcerative rash on the anterior of the middle finger, ulcerative rash over the back and umbilicus, periorbital edema with partly healed heliotrope rash, and gingival vasculopathy (Fig. 1) were noted. The gingival changes may be surrogate for an underlying vasculitis which signifies severe JDM, also supported by the poor response to high-dose corticosteroids. Therefore, the child was treated with intravenous methylprednisolone and rituximab. Over the next 5 months, the child gradually improved with complete resolution of the heliotrope rash, gingival vasculopathy, skin ulcers, and periungual rash.

Discussion

The periungual rash in this case signifies capillary changes which have been reported frequently in patients with JDM,

although the similar phenomenon of capillary dilatation, erythema, and bush-loop formation affecting the mucosal capillaries in the form of gingival vasculopathy is uncommonly recognized [1]. Oral telangiectasia have been described in both adults and children, albeit more commonly in the latter [1]. The gingival edema and hypertrophy have been described as an epiphenomenon to the capillary changes rather than a primary feature of JDM. Vasculitis forms an integral component of the Disease Activity Index score for JDM inclusive of the eyelid and periungual capillary changes along with palatal vessel dilatation [2]. Furthermore, the oral changes may mark a widespread capillary involvement in the gastrointestinal tract and intravenous steroids may be preferred for induction in these patients [3]. Gingival vasculopathy is associated with cutaneous activity with refractory disease and thus represents a possible site for assessment of disease activity and response in JDM akin to nail fold capillaroscopy [4, 5].

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Fig. 1 Gingival vasculopathy in a child with juvenile dermatomyositis

Author's contribution All authors were involved in manuscript ideation and preparation.

Declarations

Consent to participate The father has provided a signed consent to submit this article to be published. He has seen the image, text, and other materials.

Disclosures None.

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