Gottron papule-like skin changes

Dimitri Poddighe[1,3]

[1]Department of Pediatrics, ASST Melegnano e Martesana, Vizzolo Predabissi, MI, Italy
[3]Department of Pediatrics, Università degli Studi, Pavia, Italy

Correspondence to
Dr Dimitri Poddighe, dimimedpv@yahoo.it

Accepted 22 July 2017

DESCRIPTION

A 14-year-old girl came to our medical attention for persisting/recurrent fever for several months; that symptom was associated to recurrent diffuse lymphadenopathy and urticarial skin rashes (figure 1). In the previous weeks, she complained of general malaise, diffuse arthralgia and musculoskeletal pain, in addition to persistent low-grade fever. Importantly, she developed also the so-called heliotrope rash (figure 2), being pathognomonic of dermatomyositis, although blood biochemistry showed no significant alterations of creatine kinase (115 U/µL) and aldolase (12 U/L), which represent the most specific serum markers of myopathy. As regards the complete diagnostic work-up, the inflammatory parameters resulted to be increased mildly to moderately and the autoimmunity panel (anti-nuclear antibodies (ANA), extractable nuclear antigens (ENA), anti-double stranded DNA (anti-dsDNA), cytoplasmic antineutrophil cytoplasmic antibodies (ANCA), perinuclear ANCA, rheumatoid factor (RF), anti-transglutaminases antibody (anti-tTG), cold agglutinins) showed no autoantibody. Moreover, the infectious investigations included: cytomegalovirus, Epstein-Barr virus (EBV), *Toxoplasma gondii*, Parvovirus B19, Rickettsia spp, *Bartonella henselae*, *Borrelia burgdorferi*, hepatitis A virus, hepatitis B virus, hepatitis C virus; however, only EBV serology was positive and consistent with past primary infection with the development of complete serological immunity against this virus (viral capsid antigen (VCA) IgM=16.4 U/mL, VCA IgG=275 U/mL, EB nuclear antigen IgG=89.5 U/mL). Indeed, EBV-related mononucleosis was diagnosed 1 year before. In addition to the mild and unspecific musculoskeletal involvement, another particular clinical finding was the presence of Gottron's papule-like skin lesions located on the dorsal surface of the second and third toes of the left foot (figure 3). Finally, the patient was diagnosed with amyopathic juvenile dermatomyositis and, as a consequence, she was successfully treated with hydroxychloroquine and cyclosporine after an initial therapy with prednisone.

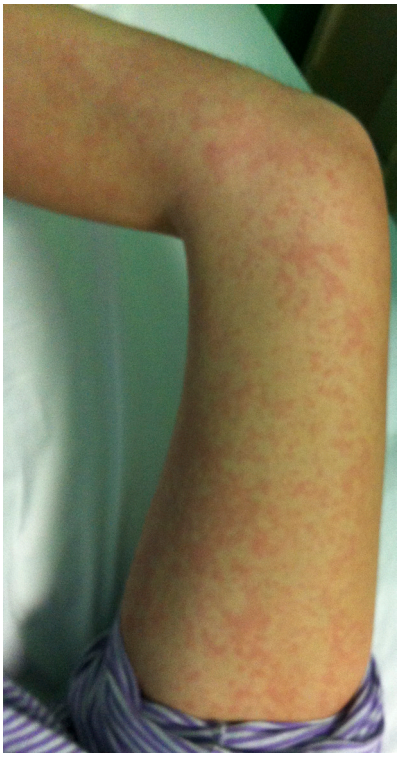

Figure 1 Urticarial rash.

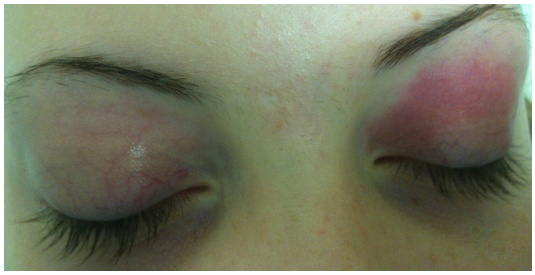

Figure 2 Heliotrope rash with eyelid oedema.

Gottron papules are symmetric changes over the extensor surfaces of joints, being characterised with erythematous and scaly plaques. Those lesions are common over the metacarpophalangeal and proximal interphalangeal joints of the hands. Gottron papules can be usually found on the extensor surfaces of the finger joints, elbows, knees or ankles, whereas the toes are affected very rarely.[1]

Here, we described a case showing Gottron papule-like skin changes as an isolated finding of this type. Moreover, this patient mainly complained of itchy urticarial rashes, as a main dermatological manifestation: indeed, a large variety of skin rashes have been described in JDM.[2] However,

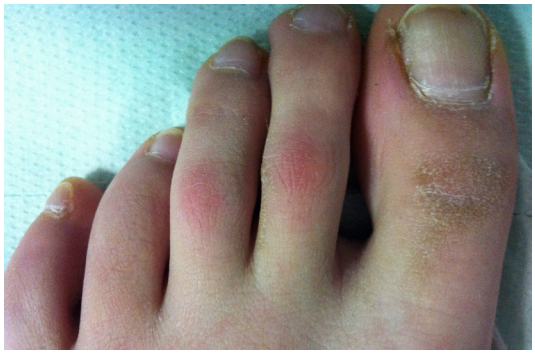

Figure 3 Gottron papule-like lesions on the toes.

To cite: Poddighe D. *BMJ Case Rep* Published Online First: [*please include* Day Month Year]. doi:10.1136/bcr-2017-221500

its association to a well-expressed heliotrope rash with eyelid oedema and to the description of several vessel changes through nail capillaroscopy (eg, microhaemorrhages, prominence of subvenous plexus, tortuosity of capillaries, bushy loop formation) supported the final diagnosis of JDM.[3]

Amyopathic juvenile dermatomyositis is a rare variant of JDM, and such a diagnosis was made after the exclusion of other immune-mediated diseases (eg, systemic lupus erythematosus, sarcoidosis, vasculitis, etc), as the corresponding diagnostic criteria were not fulfilled, and the exclusion of diseases due to infections and drugs, based on the clinical history and the specific investigations, as described previously.

Learning points

- Gottron papules must be researched in body sites that are not the usual location on extensor surfaces of metacarpophalangeal and proximal interphalangeal joints of the hands.
- Gottron papule-like skin changes can appear on the toes and show an asymmetric distribution.
- Gottron papule-like lesions have an erythematous aspect and the plaques component can be mild.

Competing interests None declared.

Patient consent Obtained from guardian.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1. Sukumaran S, Palmer T, Vijayan V. Heliotrope Rash and Gottron Papules in a Child with Juvenile Dermatomyositis. *J Pediatr* 2016;171:318–318.e1.
2 Poddighe D, De Amici M, Marseglia GL. Spontaneous (autoimmune) chronic urticaria in children: current evidences, diagnostic pitfalls and therapeutic management. *Recent Pat Inflamm Allergy Drug Discov* 2016;10(1):34–9.
3 Poddighe D, Cavagna L, Brazzelli V, *et al*. A hyper-ferritinemia syndrome evolving in recurrent macrophage activation syndrome, as an onset of amyopathic juvenile dermatomyositis: a challenging clinical case in light of the current diagnostic criteria. *Autoimmun Rev* 2014;13(11):1142–8.