Concomitant severe Kawasaki disease and pityriasis rubra pilaris in a teenager: just a coincidence?

Sir,

A previously healthy 13-year-old boy presented with fever, abdominal pain and vomiting. He was diagnosed as having a gastrointestinal infection and treated with ceftriaxone for 10 days. Two days after stopping the treatment, he complained of lower limbs mialgias, maculopapular rash, high grade remittent fever and non exudative conjunctivitis with peripheral edema. On admission into the hospital, he appeared very prostrated and febrile (40°C). Besides the previous signs, diffuse lymphadenopathy, dry mucositis and distended abdomen were also present. Laboratory examinations revealed anemia, elevated leucocyte count, ESR 85 mm/h (n.v. up to 25 mm/h), CRP 138.5 mg/L (n.v. up to 6 mg/L), elevated liver transaminase (SGPT 134 U/L, SGOT 233 U/L, GGT 188 U/L). Serum complement fractions, autoantibody profile (ANA, ENA, anti-dsDNA) were all normal or negative. Serology tests showed signs of enterovirus infection (high IgM-specific title), while tests for the other common viruses or bacteria were either negative or compatible with previous past infections. During hospitalisation, itching rapidly worsened and was poorly responsive to antihistaminic drugs. Rash became erythrodemic with eczema on face, reticular folds, limbs and abdomen. Indurate edema with keratoderma was present on hands and feet (Fig. 1). A skin biopsy, performed on day 9 of hospital stay, showed a histological picture of pityriasis rubra pilaris (PRP) (1). On day 11, the patient suddenly presented acute renal failure (serum creatinine 601 μmol/L, blood urea nitrogen 34.5 mmol/L) with oliguria and hypertension. Renal biopsy showed a picture of tubulo-interstitial nephritis (6, 7). To date, onset of PRP during KD has never been reported. PRP is an uncommon inflammatory disorder of childhood and is characterized by palmpoplantar keratoderma or erythroderma and follicular hyperkeratotic papules that coalesce into scaly erythematous plaques (1, 8, 9). KD and PRP share certain clinical features such as mucosal involvement with erythematous fissured lips, skin erythema which resolves with fine desquamation and, rarely, renal involvement (2, 8-10). All these features, along with clinical and histological PRP findings, were concomitantly present in our patient. This association might be possibly related to a pathogenetic link since both conditions are thought to be caused by an abnormal immune response or by a superantigen-mediated mechanism (10, 11).

In our case, the only positive serology test was for an enterovirus infection, already reported as trigger event in KD (12). It may be possible that an enterovirus-related toxin, absorbed through the inflamed intestinal mucosa, has stimulated local and circulating mononuclear cells which had skin and vessels as target tissues. PRP represents another possible manifestation of atypical KD. In this case, a skin biopsy should be considered in order to address the appropriate diagnosis and treatment.

F.R. PLUCHIOTT * MD
G. MARTINI, Phd
A.B. FORTINA, MD
M.C. MONTESCO *, MD
E. BENETTI, MD
F. ZULIAN, MD

Department of Paediatrics and *Institute of Pathology, University of Padua, Padua, Italy.

Address correspondence and reprint requests to: Francesco Zulian, Dipartimento di Pediatria, Università di Padova, Via Giusti 3, 35128 Padova, Italy.
E-mail: zulian@pediatria.unipd.it

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References


