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

Sirs,

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 periorbital 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 leukocyte 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 erythrodermic with eczema on face, retroauricular 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 umol/L, blood urea nitrogen 34.5 mmol/L) with oliguria and hypertension. Renal biopsy showed a picture of tubulo-interstitial nephritis. Abdominal ultrasound documented hepatosplenomegaly, gallbladder hydrops and ectatic intestinal loops. Electrocardiography evidenced an incomplete right bundle branch block while cardiac ultrasound revealed depressed ventricular function (ejection fraction 22%) and a significant right circumflex coronary artery aneurysm.

Atypical Kawasaki disease (KD) with renal insufficiency was then diagnosed (2) and therapy with intravenous immunoglobulin (IVIG) 2 gr/kg and aspirin (40 mg/kg/day) was promptly started. Renal function completely recovered after ten days of haemodialysis.

Hyperkeratotic lesions were treated with occlusive medications of betamethasone-salicylic acid ointment associated with emollient cream. Fifty days after admission the patient was discharged with improved cardiac function and skin involvement and normal renal function. PRP persisted for two months while coronary involvement normalised in six months.

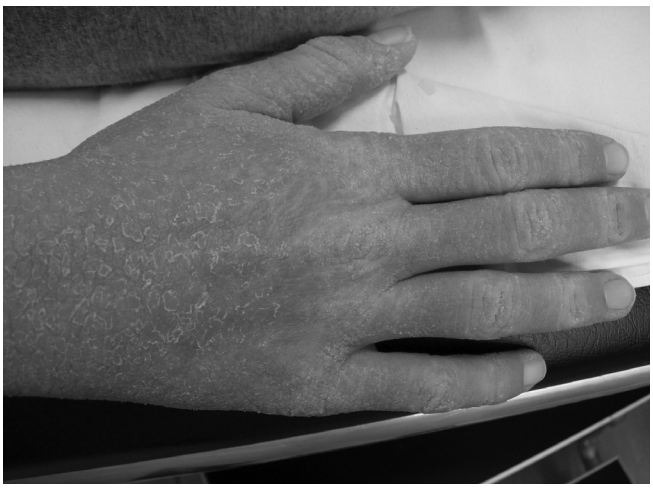

Fig. 1. Scaly erythematous plaques with hyperkeratosis and membranous desquamation of the hand. No nail abnormalities are present.

Atypical KD usually affects very young children or teenagers and is characterised by unusual clinical manifestations with high risk for misdiagnosis and increased rate of cardiac complications (3, 4).

The patient herein reported had atypical KD features, including age at presentation, systolic function depression, biliary tree involvement, acute renal failure and pityriasis rubra pilaris.

KD may cause systolic function depression in up to 20% of patients and enlarged gallbladder in over 10% (5). Renal involvement is usually characterized by proteinuria, sterile pyuria and hematuria. Acute renal failure has been reported in only 11 patients and renal biopsy, performed in 4, showed 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 characterised by palmoplantar 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 erytematous 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 a recent 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.

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