JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS MIMICKING IGA VASCULITIS

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BACKGROUND

Juvenile systemic lupus erythematosus (jSLE) is commonly presented with hematological, articular, renal and mucocutaneous manifestations. The classic rash, annular, psoriasiform lupus and chronic lesions (discoid and panniculitis), may not be present at the diagnosis, but less common forms (vasculitis) can appear mimicking Henoch–Schönlein purpura (HSP). This vasculitis is the most common in pediatric patients, being characterized by erythematous and purpuric palpable lesions without thrombocytopenia. Arthritis, abdominal or testicular pain are another characteristic manifestations. These complaints are usually mild, but may present as a severe case: nephritis, pneumonitis or central nervous system (CNS) vasculitis. The association between jSLE and HSP is infrequent.

CASE REPORT

A ten-year old girl, weighting 20 kg, was referred by the pediatric clinic to rheumatologic assessment. She started with polyarthritis and palpable purpura in both superior and inferior limbs, including palmoplantar region (Figure 1). Her first exams showed a decreased C3 complement, 24-h urinary protein 2,000 mg and high creatinine (0.75 mg/dL). The initial diagnosis HSP was made and started on 2 mg/kg prednisolone. After two months of treatment and nephritis remission failure (24-h urinary protein 4,500 mg), the child began fever, nocturnal headaches, and abdominal pain. The rheumatologist was called for evaluation. On the physical exam, the girl evidenced hepatomegaly with no other abnormalities. There was no vasculitis and arthritis in that time. Additional exams, after excluded infections causes, showed elevated erythrocyte sedimentation rate (80 mm/h), normal reactive C protein, C3 complement 41 mg/dL, C4 complement 1 mg/dL, positive ANA 1/640 speckled pattern, mild elevation on transaminases, leukocytes and dysmorphic hematuria on urinalysis, a positive double-stranded DNA (dsDNA), a positive RO and nucleosome antibodies. IV class lupus nephritis was estimated. A brain magnetic resonance assessment showed a focal area vasculitis (Figure 2). The liquor was normal. This child was hospitalized, started methylprednisolone and cyclophosphamide, developing CNS clinical and fever improvement and remission nephritis after six months.

Figure 1. Palmoplantar vasculitis and purpuric palpable lesions.

Figure 2. A single area of central nervous system vasculitis.

CONCLUSION

The report case demonstrated the relevance in considering a jSLE when vasculitis manifestations in children are presents. Atypical vasculitis areas (palmoplantar lesions) must suspect a differential diagnosis from HSP. The CSN vasculitis due to HSP often decreased C3 complement, but not C4. Finally, the hepatomegaly is not common on HPS.

KEYWORDS

Henoch–Schönlein purpura, Juvenile systemic lupus erythematosus, Vasculitis.