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CUTANEOUS POLYARTERITIS NODOSA IN A 6-YEAR-OLD CHILD WITH DOCUMENTED STREPTOCOCCAL INFECTION: CASE REPORT

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BACKGROUND

Cutaneous polyarteritis nodosa (cutaneous PAN) is a vasculitis involving small and medium sized arteries and has been associated in patients with a limited disease, without systemic features (except myalgia, arthralgia and non-erosive arthritis). It manifests with subcutaneous nodules, livedo reticularis and cutaneous ulceration, this form being associated with streptococcal infection. We are going to report a case of cutaneous PAN in a 6-year-old child with elevated antistreptolysin O (ASLO) titers.

CASE REPORT

A 6-year-old boy is evaluated for persistent fever. In the anamnesis he reported myalgias, arthralgias, fatigue. Upon physical examination there was evidence of Raynaud phenomenon, subcutaneous nodules in lower limbs and necrotic lesion in fingertips. In laboratory evaluation ANA, anti-Ro, anti-La, anti-SM, anti-cardiolipins, lupus anticoagulant, anti-beta 2 glycoprotein, anti-HIV, cytomegalovirus IgM, Epstein barr IgM, Herpes simplex IgM and ANCA were negative. There was also an increase in inflammatory tests, thrombocytosis, ASLO with titers higher than 2000 and simple urinalysis without alterations. Necrotic ulcer biopsy in fingertips demonstrated arteritis (infiltration of neutrophils, lymphocytes, eosinophils) with hypodermic vessel thrombosis. Upon the diagnosis of cutaneous PAN, he received pulse therapy with 30mg/kg of methylprednisolone for 3 days and started treatment with intravenous cyclophosphamide. Patient had an excellent response, with resolution of fever and subcutaneous nodules, without new ischemic lesions. He was discharged with prednisolone 1mg/kg, prophylactic benzathine penicillin every 21 days and a plan to complete 6 monthly infusions of cyclophosphamide in outpatient regimen.

CONCLUSION

Cutaneous PAN is rare in childhood and considered part of the spectrum of juvenile PAN and does not present specific criteria for its diagnosis. The disease should be considered in patients with fever, fatigue, myalgia, arthralgia, cutaneous infarcts, livedo reticularis and subcutaneous nodules without visceral involvement (neuropathy, hypertension, renal disease). There are several reports in the literature on the association of cutaneous PAN with streptococcal infections. Since these infections are especially common in children, research on streptococcus should be considered in patients with symptoms suggestive of PAN. The prognosis is good and the treatment is based on the severity of the disease, with mild forms responding to anti-inflammatories, topical corticosteroids and more severe or recurrent forms to systemic immunosuppressive therapy (azathioprine, mycophenolate, pulse therapy corticosteroid, cyclophosphamide, immunoglobulin). Benzathine penicillin prophylaxis appears to be beneficial and is associated with a better disease control.