Finger Necrosis as Initial Manifestation of Cutaneous Polyarteritis Nodosa in A Child: Case Report

Parmak Nekrozu ile Başvuran Bir Çocuk Olgu: Kutanöz Poliarteritis Nodoza

Cemile DURMAZ, MD, a
Nurcan CENGİZ, MD, b
Esra BASKIN, MD, c
Zekai AVCI, MD, d
Yasemin USLU, MD, b
Ali ANARAT, MD e

ABSTRACT Polyarteritis nodosa (PAN) is a necrotizing vasculitis involving small and medium-sized arteries, that most often affects the kidneys, heart, and liver, but can involve any organ system. A cutaneous form of PAN (CPAN) without visceral involvement that follows a benign but often chronic course has been described. Skin findings of CPAN are maculopapular rash, subcutaneous nodules, livedoid vasculitis or erythematous patchy rashes. In this report, we describe a 10-year-old girl with CPAN with bilateral necrotic lesions on her toes. Her erythrocyte sedimentation rate and C-reactive protein level were elevated. Treatment was started with a heparin infusion. The presence of thromboembolic disease was ruled out with normal coagulation tests and normal Doppler ultrasonography of lower extremities and heparin infusion was withheld in three days. After skin biopsy was done, intravenous pulse methylprednisolone therapy was prescribed. Marked improvement of her skin lesions occurred after methylprednisolone teratment, and the patient was discharged on a regimen of oral prednisone and aspirin. Finger necrosis as in the present case, also should be considered as a clinical sign for CPAN.

Key Words: Vasculitis, polyarteritis nodosa, child


Anahtar Kelimeler: Vaskülti, poliarteritis nodoza, çocuk


Polyarteritis nodosa (PAN) is a necrotizing vasculitis involving small and medium-sized arteries, that most often affects the kidneys, heart, and liver, but can affect any organ system.1 A cutaneous form of PAN (CPAN) without visceral involvement that follows a benign but often chronic course also has been described. Skin findings in CPAN include maculopapular rash, subcutaneous nodules, livedoid vasculitis, panniculitis, ischemic finger lesions, or erythematous patchy rashes. Here, we describe a patient with CPAN with finger necrosis as initial manifestation.

Copyright © 2010 by Türkiye Klinikleri
CASE REPORT

A 10-year-old girl was admitted to our pediatric unit with purple coloured skin lesions. She had an upper respiratory infection 15 days earlier. The patient had no abdominal pain, fever, change in urine color or hypertension. Her medical and family histories were unremarkable. On physical examination, she had bilateral blue-purple coloured toes. There was severe necrotic appearance on the distal 1/3 of both feet (Figure 1). She also had erythematous lesions on her nose, ears, hands, and gluteal region. Both feet were slightly swollen and tender. Her blood pressure was 110/70 mmHg all peripheral pulses were palpable. Results of the rest of the medical examination were normal. Laboratory analyses revealed the following: hematocrit, 42%; white blood cell count, 13800/mm³ with 76% neutrophils and 24% lymphocytes; platelet count, 400 000/mm³; erythrocyte sedimentation rate, 65 mm/h; and C-reactive protein, 40 mg/L. Urinalysis was normal. Results of tests for electrolyte concentrations, renal and liver function tests, viral serology for Epstein-Barr virus, cytomegalovirus, hepatitis A, B and C, Salmonella and Brucella agglutination tests, coagulation tests of prothrombin time, partial thromboplastin time, fibrinogen, D-Dimer, protein S and C, antitrombin-II-I, antinuclear antibody, antiDNA, rheumatoid factor, antestreptolysin O titer, anticardiolipin antibody, anti-neutrophilic cytoplasmic antibodies (perinuclear, cytoplasmic), C3 and C4 complement components were either normal or negative. Factor 5 Leiden mutation and prothrombin polymorphism analysis were normal. Doppler ultrasonography of lower extremities was normal. A skin biopsy from right foot showed several medium-sized muscular arteries demonstrating perivascular eosinophilic and neutrophilic infiltrates, fibrinoid necrosis, and vascular destruction (Figure 2). Also, IgM and IgG deposition on the arterial wall was seen (Figure 3).

Treatment was started with a heparin infusion (20 U/kg/hour). The presence of thromboembolic disease was ruled out with normal coagulation tests and normal doppler ultrasonography of lower extremities, heparin infusion was withheld in three days. After a diagnosis of CPAN was made, the patient received an intravenous high-dose methyl prednisolone infusion (30 mg/kg/day) for three days. Oral prednisone (1 mg/kg/day) and aspirin 100 mg/day started. Marked improvement of her skin lesions occurred after several days, and two weeks later the patient was discharged on a regimen of oral predni-
sone (1 mg/kg/day) and aspirin. Oral prednisone and aspirin treatment was continued four months. On 6-months follow-up, the patient's clinical findings were normal, with no apparent skin lesions.

**DISCUSSION**

Cutaneous PAN in children is extremely rare. The first case was reported by Velbov in 1980. It is a rare form of vasculitis that appears to be limited primarily to the skin, muscles, and joints. In contrast to the systemic form of the disease, CPAN is characterized by an absence of visceral lesions and a relapsing, but benign course.1

The clinical manifestations are painful cutaneous nodules localized mainly on the lower extremities, although other lesions such as livedo reticularis, ulcers, maculopapular rash, erythematous patchy rashes, or ischemic finger lesions may be seen.2,4 Our patient presented with ischemic lesions on her feet, which is a rare initial manifestation of CPAN.

Laboratory tests have usually been unremarkable, except for elevated ESRs, as was the case in our patient. It has been reported that while an elevated ESR in systemic PAN is a bad prognostic sign, elevated ESRs in cases of cutaneous PAN have been followed by a recurrent, but benign course.5 The CRP also was elevated in our patient, as was true in a previously reported adult patient with acral necrosis on both hands as the initial manifestation of CPAN.6

The diagnosis of CPAN is made clinically and with a biopsy examination of the affected skin by demonstrating fibrinoid necrosis of the small and medium-sized arteries, similar to those in our patient.

The etiology of CPAN is unknown. Based on the demonstration of IgM and C3 deposits in lesional biopsy specimens and the detection of circulating immune complexes, immunologic mechanisms might play a potential role in CPAN as a systemic disease.7 IgM deposits were demonstrated in a skin biopsy of our patient. According to some but not all authors, cutaneous and systemic PAN in children often occur after an upper respiratory tract illness (URTI) or documented streptococcal infection. In a large series of patients with CPAN, 42% of patients had an associated URTI at the start or with relapses of the disease. These patients described a history of URTI or had positive cultures or serology for streptococci.5 Associations with streptococcal infections also suggest immunocomplex mediation. Our patient had a history of URTI, but we could not find a high antistreptolysin O titer. In children, a possible association between CPAN and hepatitis viruses B and C has been noted.7 In our patient, results of serologic tests for hepatitis B and C were negative.

Aspirin, prednisolone, and methotrexate, alone or in combination, have relieved acute exacerbations, but some patients require high-dose corticosteroids, and slow tapering is usually required.3,5 Antibiotic treatment may be needed in patients with documented streptococcal or other bacterial infections.5 Some authors recommend penicillin prophylaxis in patients with documented streptococcal disease.5,8 Our patient was treated with heparin infusion for three days, and thereafter intravenous high-dose methyl prednisolone was started at 30 mg/kg/day for three days and tapered to oral prednisolone in combination with aspirin. We observed marked clinical improvement with high-dose methyl prednisolone and she has had no recurrences during 6 months follow-up.

Finger necrosis as in the present case, should be considered as a clinical sign for CPAN. We suggest that the high-dose methyl prednisolone is useful in the treatment of patients with dramatic finger necrosis in CPAN.

**REFERENCES**