

Cutaneous Polyarteritis Nodosa Presenting with Digital Gangrene

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Abstract

Cutaneous polyarteritis nodosa is a vasculitis with characteristic manifestations like tender subcutaneous nodules, livedo reticularis, cutaneous ulcers and necrosis. Diagnosis requires histopathologic evidence of necrotizing inflammation of the medium and small arteries. We report a six year old girl with cutaneous PAN with gangrenous changes of the fingertips which responded to methylprednisolone.

Key words: Cutaneous polyarteritis nodosa, subcutaneous nodules, Vasculitis, Gangrene, Steroids

Introduction

Cutaneous Polyarteritis nodosa (CPAN) is a vasculitis designated for the cutaneous limited form of Polyarteritis nodosa (PAN) affecting the small and medium vessels of the dermis without any systemic involvement^{1,2}. It affects all ages but uncommon in children^{1,3}. Lindberg first recognized the CPAN and approximately 140 cases have been reported in the literature so far^{3,4}. The characteristic manifestations include tender subcutaneous nodules, livedo reticularis and ulceration, mostly localized to the lower extremity^{1,4,5}. The definitive diagnosis requires clinical and histopathologic evidence of necrotizing inflammation of the medium and small arteries^{2,4}. CPAN has a chronic, relapsing course which is steroid responsive^{1,2,4}. We report a six year old girl with cutaneous PAN, who developed gangrenous changes of the fingertips which responded to methylprednisolone.

The Case

A six year old girl presented with fever, joint pains and skin lesions since 1 week. On examination, she had knee and ankle joint synovitis of dorsum of hand and feet. Skin examination revealed multiple tender subcutaneous nodules in all the limbs with lower limb preponderance (Fig 1A,B). Systemic and ophthalmological examination was remarkable. Investigations revealed Hemoglobin 9.9g/dL, WBC 21800/ μ L (Neutrophils 68%, lymphocytes 27%), platelets 6.1 lakhs/ μ L, erythrocyte sedimentation rate 75mm/hr, Antistreptolysin O (ASLO) positive >200 IU/mL (Normal=0-200IU/mL), Serum Ferritin 167U/L (0-120), C-reactive protein 14.1 mg/dL (normal range <0.5) and normal peripheral blood smear. Her renal function tests, liver function tests

and bone marrow aspiration study were normal. Stool analysis showed negative for occult blood. The blood and urine cultures were sterile. Antibodies against hepatitis B, hepatitis C and human immunodeficiency virus were not detected. Her mantoux test, rheumatoid factor, ANA and ANCA by IFA were negative. Chest X-ray, abdominal ultrasonography and echocardiography of the heart were normal. Skin biopsy from subcutaneous nodule showed unremarkable epidermis. Dermis showed dilated capillaries with sparse perivascular lymphocytic infiltrates and the deep dermis and subcutis show blood vessel wall infiltrated by lymphocytes, eosinophils and neutrophils suggestive of medium vessel vasculitis with IF being negative.

The clinicopathological presentation was consistent with CPAN and she was treated with prednisolone and methotrexate. There was a complete resolution of symptoms over a period of 3 months. At the fourth month, she presented with bluish black discoloration of the right ring and little fingertips with recurrence painful subcutaneous nodules (Fig 1 C). Radial pulses were symmetric and normal. Allen's test was normal. Her symptoms completely resolved in two weeks time after pulse methyl prednisolone (Fig 1 D). On discharge child was put on oral steroid therapy and advised regular follow up. Follow up for six months was unremarkable.

Discussion

Polyarteritis nodosa is a rare vasculitis in childhood⁴. Children were classified into three forms: infantile, cutaneous, and systemic forms⁴. Cutaneous PAN is recognized as a separate entity without any

diagnostic criteria and is classically devoid of systemic involvement of the liver, kidney or heart. A new diagnostic criterion has been designed by Nakamura et al¹. Our child satisfies all the criterias. The characteristic manifestations include tender subcutaneous nodules, livedo reticularis and ulceration, mostly localized to the lower extremity^{1,4,5}. Extracutaneous symptoms such as peripheral neuropathy, myalgia, and arthralgia are rare^{1,5}. A typical burst pattern of irregularly shaped livedo reticularis around an ulcer is highly suggestive of CPAN⁵. CPAN usually involves lower limbs (80-97%), followed by the arms(33%), and the trunk(8%). Most of them had subcutaneous nodules (86%), followed by livedo reticularis (45%), purpura (45%) and ulcers (23%)⁵.

The most common inciting agent is Group A β hemolytic Streptococcus⁵. Testing for group A streptococcal infection was positive in 86.2% of cases⁴. Fathalla et al reported four children of CPAN with elevated streptococcal enzymes in three of them⁶. Our case typically presented with cutaneous manifestations without systemic involvement, also ASLO positive and the diagnosis was confirmed by skin biopsy. Her initial episode was treated with steroids and methotrexate to which she responded well initially. The disease relapsed at the fourth month with severe ischemic changes of the fingertips which promptly responded to methylprednisolone. Choi et al reported CPAN with digital gangrene in a 34 year old woman and showed improvement of skin lesions with corticosteroid & alprostadil but the finger required amputation⁷. However, Stussi et al described a case of CPAN with digital necrosis in a 33 year old woman, she responded for prostaglandin, calcium channel blocker, and corticosteroid⁸.



Fig 1: a) Multiple tender subcutaneous nodules in the Palm.
b) Multiple tender subcutaneous nodules in the Foot



Fig 2: a) Bluish black discoloration of the right ring and little fingertips
b) Resolved gangrenous changes after methylprednisolone therapy

Cutaneous PAN can be challenging to the clinician to diagnose and manage⁹. Differential diagnosis of painful subcutaneous nodules include CPAN, erythema nodosum, Systemic PAN and other vasculitis like HSP⁵. CPAN should be differentiated from systemic polyarteritis nodosa because both of them have different clinical course and management^{2,3}. The extra-cutaneous manifestations of peripheral neuropathy and myalgia in CPAN occur only adjacent to the cutaneous lesions, while they may be disparate in systemic PAN^{1,5}. Even though skin manifestations commonly seen in the lower limbs in HSP, palpable purpuras are non tender unlike in CPAN.

Histopathologic evidence of necrotizing inflammation of the medium and small-sized arteries is characteristic in CPAN^{4,5,9}. Blood counts, ESR, liver and renal function tests, cryoglobulins, anti-nuclear antibody (ANA), anti-neutrophil cytoplasmic antibody (ANCA), rheumatoid factor, and complement levels are necessary to exclude other causes of vasculitis and systemic PAN⁵. Most of the laboratory findings are non-specific¹⁰. CPAN runs chronic course with remissions, relapses, with a favorable prognosis⁵. There is no evidence regarding the CPAN progression to systemic

PAN⁵. Corticosteroids remain the mainstay of treatment for CPAN². Autoamputations of fingers and toes due to gangrene may be because of late presentation. However our child gangrene responded to Methyl prednisolone. Even though our child initially responded to prednisolone, during the follow up developed gangrenous changes of finger tips which responded to methyl prednisolone.

There is no relationship between the age of onset and the disease severity in CPAN⁵. Out of all 79 cases of CPAN systemic PAN did not develop in any patient¹⁰. Similarly none of the 22 cases had progression in a study by Nakamura et al¹.

Conclusion

CPAN should be considered in a child who presents with tender subcutaneous nodules, livedo reticularis along with constitutional symptoms like fever, myalgias, and arthralgias/arthritis. The diagnosis is purely clinic-histopathologic correlation. Most children respond to corticosteroids. Even though ischemic and gangrenous changes are rare manifestations of CPAN, early notifications help in the active management to prevent amputations and consequences.

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