

Cutaneous Polyarteritis Nodosa: A case report

Harish J, Manjunath M N and Chaithanya C Nair

Abstract

Cutaneous polyarteritis nodosa is a rare vasculitis of childhood relating to small-to-medium-sized arteries. Its etiology is unknown. Clinical manifestations include tender subcutaneous nodules, livedo reticularis, cutaneous ulcers and necrosis. Although it is distinct from systemic polyarteritis nodosa in that it lacks significant internal organ involvement, extra-cutaneous manifestations may be evident. The diagnosis is by skin biopsy as there is no other specific serological tests and characteristic pathologic feature is a leukocytoclastic vasculitis in the small to medium-sized arterioles of the dermis with or without fibrinoid necrosis. Mild cases may resolve with nonsteroidal anti-inflammatory drugs. If more severe, treatment with systemic corticosteroids generally achieves adequate response. We report a 10 year old girl with cutaneous PAN, who presented to us with arthralgia and swelling of left knee joint and both ankle joints and fever, with multiple tender subcutaneous nodules on both upper and lower limbs.

Keywords: Cutaneous polyarteritis nodosa, subcutaneous nodules, vasculitis.

Abbreviations: cPAN- Cutaneous polyarteritis nodosa, PSM- pansystolic murmur. CRP- c reactive protein, ASLO- anti streptococcal lysin O

Polyarteritis nodosa (PAN) is a rare vasculitis in childhood. Since first described by Kussmaul and Maier in 1866¹, there have been approximately 140 pediatric case reports in the literature. Traditionally, children were classified as having one of three forms: infantile, cutaneous, and systemic. Infantile PAN is now recognized as a severe form of Kawasaki disease. Criteria for a diagnosis of systemic PAN in childhood have been proposed but not validated².

Cutaneous PAN (cPAN) is recognized as a separate entity but there are no diagnostic criteria for cPAN². cPAN is characterized by disease affecting the skin with no major organ system involvement. The cutaneous symptoms are similar to systemic PAN and mild fever, muscle, joint, and peripheral nervous system involvement may also occur. Fever, rash, and musculoskeletal symptoms are common in children and cPAN needs to be differentiated from other diagnostic entities. Definitive diagnosis is by histopathologic evidence of necrotizing inflammation of the medium and small-sized arteries. There is a paucity of knowledge of the spectrum of clinical presentation and management of children with cPAN. Here we describe a case of cPAN and summarize the clinical manifestations, laboratory data and treatment regimens of our patient.

Case report

This 10 year old female adolescent presented with pain in both the elbow joints followed by pain in the left knee joint and both the ankle joints in a course of 8 days and fever for the past 2 days. On admission her vitals were stable, both the elbow joints were tender and the knee and ankle joints were swollen and tender. She had multiple subcutaneous nodules over extensor aspect of both her forearms, both her tibial shins and few on her thighs. Systemic examination showed presence of a soft PSM of

grade 2 intensity over left sternal edge. Blood investigations showed leucocytosis, elevated CRP, elevated ASLO titres and 2D echo revealed a mild tricuspid regurgitation. An initial diagnosis of acute rheumatic fever was made and child was started on penicillin and Aspirin. But child continued to have excruciating arthralgia and hence a rheumatologist opinion was taken. Child was advised a skin biopsy from the nodular lesions which showed small and medium vessel vasculitis suggestive of cutaneous polyarteritis nodosa.

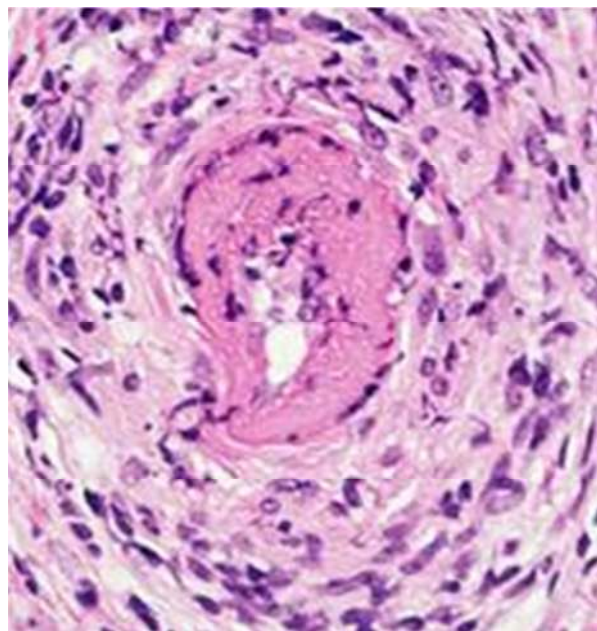


Fig 1 showing segmental fibrinoid necrosis with inflammatory infiltrates of small artery.

Hence she was stopped with aspirin therapy, given pulse therapy with methyl prednisolone and continued with penicillin therapy. Her arthralgia subsided within a day of pulse therapy and the subcutaneous nodules gradually disappeared. On

discharge child was put on oral steroid therapy and penicillin prophylaxis and advised regular follow up.

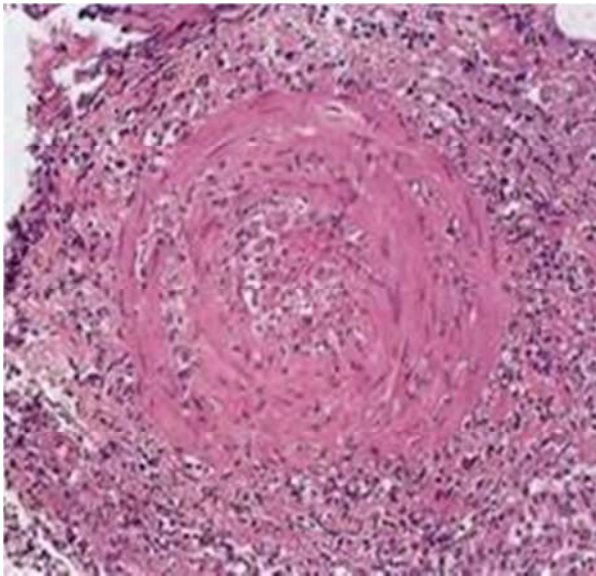


Fig 2 showing leukocystoclastic vasculitis with fragmentation of neutrophils in and around blood vessels.

Discussion

cPAN is not common in the pediatric population with approximately 140 cases reported in the literature. Disease is limited to skin, joints, and muscles in the majority with a minority having nerve involvement. Constitutional symptoms are common. Most children have a chronic and relapsing benign course.

The precise etiology of cPAN remains to be unknown. However, an immune mediated mechanism has been postulated. Several infectious and noninfectious conditions have been associated both to initiation and relapse of the disease^{3,4,5}. Among them, streptococcal infection has been commonly implicated^{6,7}. Although some evidence of streptococcal infection as an initiating factor for cPAN is present, caution must be exercised when interpreting elevations in the serologic markers of streptococcal infection in the absence of an appropriate clinical presentation.

Cutaneous and systemic PAN share the same histopathologic features of necrotizing arteritis of small and medium sized vessels. Kussmaul and Meier described the first case of systemic PAN in 1866¹. Early reports^{8,9} confirm that cPAN is a separate entity to systemic PAN. We have limited our definition of cPAN to disease affecting the skin, muscle, joints, and peripheral nervous system, with corresponding biopsy confirmation. Any evidence of visceral involvement, either clinically (central nervous system, pulmonary, cardiac, gastrointestinal, or renal), radiographically (abnormal angiography), or by histology (visceral biopsy) were classified as systemic PAN. Nakamura et al¹⁰ proposed further restriction of the definition of cutaneous PAN in that any extracutaneous involvement such as peripheral neuropathy and myalgias must

be limited to the same area as skin lesions. Systemic PAN and cPAN appear to be fairly distinct entities on a clinical continuum. There are only 5 reported cases of cPAN evolving into systemic PAN^{11,12}.

On review of treatment regimens reported in the literature, most children respond to corticosteroids. Penicillin should be considered in children with increased ASO titres^{13,14}. Recent case series report success with low-dose methotrexate, cyclophosphamide, intravenous immunoglobulin, and biologic therapies^{15,16}.

In summary, cPAN can be challenging to diagnose and manage. A diagnosis of cPAN should be considered in a child with fever, tender subcutaneous nodules, livido reticularis, and arthralgias/arthritis. Most children respond to corticosteroids and have a benign course, but some children may be corticosteroid dependent or corticosteroid resistant, necessitating other immunosuppressive agents including DMARDs and biologic therapy. Multicentre pediatric vasculitis disease registries are necessary to inform development and standardization of best clinical practice for childhood cPAN.

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Competing Interests

None declared

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