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Case Report

A Case Report of Childhood Polyarteritis Nodosa: A Rare Vasculitic Disorder with Multisystem Involvement

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Abstract

Background: Childhood polyarteritis nodosa (cPAN) is a rare systemic necrotizing vasculitis affecting small- and medium-sized arteries. Its heterogeneous clinical manifestations often delay diagnosis, yet early recognition is essential to prevent organ damage. According to the EULAR/PRINTO/PRES criteria, diagnosis requires evidence of necrotizing vasculitis in medium- or small-sized arteries or characteristic clinical features such as skin involvement, myalgia, hypertension, or peripheral neuropathy. This case adds to the limited literature by presenting a severe multisystem form of cPAN in a young child who achieved favorable outcomes with timely corticosteroid therapy.

Case Presentation: A 3-year-old girl presented with a 6-month history of recurrent fever, painful subcutaneous nodules, pruritic rashes, digital gangrene, myalgia, arthralgia, proteinuria, hematuria, and hypertension. Histopathologic examination of skin and subcutaneous nodules demonstrated necrotizing vasculitis of medium-sized arteries, fulfilling the EULAR/PRINTO/PRES classification criteria for cPAN. Treatment with oral prednisolone (1 mg/kg/day) and supportive care led to rapid improvement, including regression of skin lesions, normalization of inflammatory markers, resolution of renal involvement, and sustained remission during follow-up.

Conclusion: This case highlights the need for high clinical suspicion of cPAN in children presenting with multisystem symptoms and characteristic cutaneous findings. Early diagnosis supported by established criteria and prompt corticosteroid therapy, combined with multidisciplinary care, can significantly improve outcomes and reduce long-term morbidity.

Keywords: Polyarteritis nodosa; childhood; vasculitis; prednisolone; case report

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Introduction

Polyarteritis nodosa (PAN) is a rare and potentially fatal systemic necrotizing vasculitis that primarily affects medium-sized arteries, though small arteries may also be involved. [1] Childhood polyarteritis nodosa (cPAN) tends to present with a more variable and often rapidly progressive clinical course. Its exact prevalence remains unknown, but it shows a male predominance with a male-to-female ratio of approximately 2:1. [2]

While the majority of cases are idiopathic, several secondary causes have been identified. These include infections such as hepatitis B and C, Klebsiella species, and other bacterial pathogens, as well as associations with malignancies. Furthermore, cPAN has been linked to autoimmune diseases, including rheumatoid arthritis and Sjögren's syndrome. [2, 3]

The underlying pathophysiology is believed to involve immune complex-mediated inflammation of medium-sized vessels, leading to vascular ischemia and tissue infarction. This results in multisystem involvement, most commonly affecting the kidneys while typically sparing the lungs. Other organ systems frequently involved include the skin, central nervous system, heart, gastrointestinal tract, and musculoskeletal system.

The diagnosis of PAN is confirmed by demonstrating vascular involvement through tissue biopsy and/or angiographic evidence of aneurysms, stenoses, or occlusions. Prognosis varies depending on disease severity and organ involvement. However, childhood PAN generally carries a lower mortality risk compared to the adult form. [4]

Case Presentation

A 3-year-old girl presented with a 6-month history of recurrent low-grade fever, painful subcutaneous swellings, and widespread pruritic rashes. These symptoms were accompanied by progressive myalgia, arthralgia, generalized weakness, anorexia, weight loss, and eventual inability to walk due to pain. There was no joint swelling. Symptoms began one week after a blood transfusion administered during an acute febrile illness at a peripheral hospital.

On examination, she was in painful distress and severely undernourished (BMI 11.4 kg/m^2; weight 8 kg, 57% of expected; length 84 cm, z-score < −2 SD). Cutaneous findings included livedo reticularis (as shown in Figure 1) and generalized maculopapular rashes, multiple tender, hyperpigmented subcutaneous nodules on the trunk and limbs (as shown in Figure 2), with erythematous lesions on the palms and soles. Dry gangrene affected the distal pulp, nail, and nail beds of the right thumb and index finger and the left little finger (as shown in Figure 3).

Figure 1: Livedo reticularis on the dorsum of the foot in a child with childhood polyarteritis nodosa.

Figure 2: Subcutaneous nodules with reticulated livedo reticularis on the posterior thoracolumbar region.

Figure 3: Dry gangrene of the distal phalanges of the right thumb and index finger and left little finger at presentation.

Musculoskeletal examination revealed marked diffuse myalgia and arthralgia limiting standing and ambulation. Her blood pressure was elevated at 130/90 mmHg (>95th percentile for age), with renal involvement evidenced by proteinuria and hematuria.

Skin punch biopsy showed leukocytoclastic vasculitis with fibrinoid necrosis of small dermal vessels. Deep biopsy of a subcutaneous nodule demonstrated necrotizing vasculitis of medium-sized arteries with infiltration by neutrophils and lymphocytes, internal elastic lamina disruption, and surrounding chronic inflammation—findings consistent with childhood polyarteritis nodosa. A chest radiograph was normal. Relevant laboratory investigations are summarized in Table 1.

Table 1: Laboratory Findings of a 3-Year-Old with Childhood Polyarteritis Nodosa

Parameter	Patient Value	Reference Range
Urinalysis (Initial & Repeat)	Proteinuria ++, Hematuria ++	Negative for protein and blood
White Blood Cell Count (WBC)	19.9 × 10^3/Ml	4.0 – 11.0 × 10^3/µL
Neutrophils	72.4%	35% – 55%
Lymphocytes	10.5%	35% – 55%
Monocytes	10.7%	2% – 8%
Hematocrit	29.7%	Male: 35% – 40%
Platelet Count	377 × 10^3/Ml	150 – 400 × 10^3/µL
Erythrocyte Sedimentation Rate (ESR)	130 mm/hr	< 15–20 mm/hr
Sodium (Na^+)	137 mmol/L	135 – 145 mmol/L
Potassium (K^+)	3.8 mmol/L	3.5 – 5.0 mmol/L
Bicarbonate (HCO_3^-)	22 mmol/L	22 – 28 mmol/L
Urea	2.6 mmol/L	1.8 – 6.5 mmol/L
Creatinine	35 µmol/L	27 – 62 µmol/L
Hepatitis B Surface Antigen	Non-reactive	Negative
Anti-HCV Antibody	Non-reactive	Negative
HIV Screening	Non-reactive	Negative
ANA	Negative	Negative
Blood culture	No culture yielded	

Abbreviations: ESR = erythrocyte sedimentation rate; HCO_3^- = bicarbonate; Na^+ = sodium; K^+ = potassium; HCV = hepatitis C virus. ANA = antinuclear antibodies

Table I: Laboratory Findings of a 3-Year-Old with Childhood Polyarteritis Nodosa

The patient's laboratory results (Table 1) demonstrated systemic inflammation and renal involvement, evidenced by leukocytosis, neutrophil predominance, elevated ESR, and proteinuria with hematuria. Viral serologies were negative.

Management and Outcome

The patient was started on oral prednisolone (1 mg/kg/day), loratadine, short-course intravenous followed by oral amoxicillin–clavulanate, and analgesics. Within 24–36 hours, she showed marked improvement: pain diminished, she resumed walking and playing, and the skin lesions began to regress. By day 5, gangrenous areas showed clear demarcation, and by day 12, auto-amputation of the distal left little finger occurred, with fibrosis of the right index and middle finger nail beds (as shown in Figure 4).

Figure 4: Auto-amputation and healed fibrotic changes of the digits following corticosteroid therapy.

By the end of the second week, her blood pressure improved to 100/60 mmHg, and urinalysis revealed only mild proteinuria. She was discharged on tapering prednisolone. At 4-week follow-up, blood pressure had normalized, urinalysis was negative for hematuria and proteinuria, leukocyte count was $5.8 \times 10^3/\mu L$, and ESR decreased to 12 mm/hr. She remained clinically stable on tapering corticosteroids, with remission sustained for six months post-treatment. Biweekly follow-up continued to monitor for relapse. A timeline of clinical events is summarized in Table 2. Figure 4 above shows the healed fibrotic changes.

Table 2: Timeline of Clinical Events, Interventions, and Outcomes in a 3-Year-Old with Childhood Polyarteritis Nodosa

Time	Clinical Events	Interventions	Outcomes
~6 months before presentation	Recurrent intermittent fever, painful nodules, pruritic rashes, arthralgia, weight loss	Supportive care; blood transfusion after febrile illness	No sustained improvement
At presentation	Fever, skin nodules, livedo reticularis, digital gangrene, hypertension, proteinuria, hematuria	Admission, diagnostic workup, biopsies	Diagnosis of childhood PAN
Day 1–2	Severe myalgia/arthralgia, distress	Prednisolone 1 mg/kg/day, loratadine, IV antibiotics, IV ketorolac	Marked symptom relief, able to walk/play
Day 5–12	Regression of nodules, improved systemic signs	Continued steroids, switch antibiotics to oral, NSAIDs	Auto-amputation of little finger
Week 2	BP 100/60 mmHg, proteinuria (+)	Prednisolone tapered to 0.5 mg/kg/day	Stable clinical state
Week 4	BP normalized (88/56 mmHg), ESR 12 mm/hr, urinalysis clear	Tapering alternate-day steroids	Sustained remission

Ongoing follow-up	No relapses	Continued tapering corticosteroids	Stable clinical remission
Abbreviations: BP = blood pressure; ESR = erythrocyte sedimentation rate; IV = intravenous; NSAIDs = nonsteroidal anti-inflammatory drugs; PAN = polyarteritis nodosa.			

The timeline of clinical progression, interventions, and outcomes in the patient with childhood polyarteritis nodosa (cPAN) is presented in **Table 2.** It demonstrates the sequence of systemic involvement, diagnostic confirmation, and favorable therapeutic response following corticosteroid therapy.

Discussion

Pathophysiology of Pediatric Polyarteritis Nodosa

Classic polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis that predominantly affects small- and medium-sized muscular arteries. It is characterized by segmental stenosis and aneurysmal nodules along the vessel walls, leading to downstream tissue ischemia and multiorgan involvement. [1,4] The pathogenesis in childhood is poorly understood but is thought to involve post-infectious autoimmune mechanisms triggered by infections such as Group A Streptococcus, hepatitis B virus (HBV), Epstein-Barr virus, cytomegalovirus, parvovirus B19, and Mycobacterium tuberculosis. [1,2,4] Genetic and autoinflammatory predispositions, including familial Mediterranean fever, have also been implicated. [1, 5, 6]

Epidemiology and Unusual Presentation

Childhood PAN (cPAN) is rare, with a male predilection (male-to-female ratio ~2:1) and median age at presentation around 9 years. [5] Our patient, a 3-year-old girl, represents an atypical demographic in terms of age and sex. Early-onset cases may present diagnostic challenges due to nonspecific prodromal symptoms such as fever, malaise, weight loss, anorexia, and myalgia, which can mimic infections or post-transfusion complications. [1, 5]

Clinical Presentation and Comparison with Other Pediatric Cases

The clinical manifestations of PAN are heterogeneous and correspond to the distribution and severity of vascular inflammation. Renal involvement, commonly presenting as proteinuria and hematuria without glomerulonephritis, is frequent, and hypertension may develop secondary to renal artery involvement. [1, 4, 6] Pulmonary sparing distinguishes PAN from other vasculitides, such as microscopic polyangiitis. [1, 4, 6]

Cutaneous involvement occurs in approximately 40% of pediatric cases and may include painful subcutaneous nodules, livedo reticularis, purpura, hyperpigmentation, digital ischemia, ulcers, and gangrene in advanced disease. [4, 5] Musculoskeletal complaints are common, with myalgias and arthralgias in 50% of patients. Gastrointestinal and cardiac manifestations vary in frequency, while neurological involvement is uncommon (<10%), manifesting as transient ischemic episodes, strokes, or cerebral arteritis in late stages. [1, 4, 5]

The clinical manifestations in this case reflect the heterogeneous nature of the disease, with multisystem involvement including the skin, musculoskeletal system (arthralgia, myalgia), and cardiovascular system, with hypertension and renal involvement evidenced by proteinuria and haematuria. Importantly, the pulmonary system remained unaffected, consistent with the classical pattern of PAN. [7, 8]

In resource-limited settings, clinical presentations of pediatric PAN have been reported with delayed diagnosis and variable organ involvement. Mondal et al. conducted a multicenter prospective study from eastern India and reported that cutaneous, renal, and musculoskeletal manifestations were the most

common features, reflecting challenges in early recognition and intervention. [9] Our patient's presentation aligns closely with these findings, highlighting the relevance of cPAN patterns in low- and middle-income countries.

Diagnostic Challenges

Diagnosis of cPAN relies on clinical features supported by histopathology and/or angiography. Biopsies of affected tissue (skin or muscle) often reveal necrotizing inflammation of small- and medium-sized arteries with fibrinoid necrosis. [1, 6] Angiography can demonstrate microaneurysms, stenoses, or occlusions in mesenteric or renal arteries. [1, 6] However, in our patient, angiographic studies were not feasible due to **resource constraints,** which is acknowledged as a limitation. Diagnosis was therefore confirmed histologically via skin and subcutaneous nodule biopsy, demonstrating leukocytoclastic vasculitis and necrotizing arteritis.

The EULAR/PRINTO/PRES classification criteria for cPAN require histologic or angiographic evidence plus at least one clinical feature (skin involvement, myalgia/muscle tenderness, hypertension, peripheral neuropathy, or renal involvement). [6] Our patient met these criteria with skin, renal, musculoskeletal involvement, and hypertension, despite the absence of angiography or peripheral neuropathy. Additional laboratory findings (leukocytosis, elevated ESR, hematuria, proteinuria, and negative viral serologies) further supported the diagnosis.

Differential diagnoses considered included Henoch-Schönlein purpura, Kawasaki disease, and microscopic polyangiitis. The absence of pulmonary involvement, sparing of glomeruli, and presence of cutaneous aneurysms and digital ischemia favored PAN. [1, 4]

Management and Response

Treatment of cPAN is guided by disease severity and organ involvement. First-line therapy involves high-dose corticosteroids (oral prednisolone 1–2 mg/kg/day or intravenous methylprednisolone 30 mg/kg/day for severe disease), followed by gradual tapering. [1, 4, 7] Adjunctive immunosuppressive agents—cyclophosphamide, azathioprine, methotrexate, or mycophenolate mofetil—are indicated in organ-threatening or refractory cases, while biologic agents (rituximab, anti-TNF therapies) may be considered for resistant disease. [11–13]

Our patient received **corticosteroid monotherapy** with supportive care, including NSAIDs and empiric antibiotics. She demonstrated rapid improvement, normalization of inflammatory markers, resolution of renal and blood pressure abnormalities, regression of skin lesions, and auto-amputation of affected digits. This aligns with previous reports indicating that steroid monotherapy can induce remission in mild-to-moderate cPAN, particularly when treatment is initiated early. [5,10]

Prognostic Implications

Untreated or severe PAN may lead to skin ulceration, chronic kidney disease, gastrointestinal perforation, stroke, or aneurysm rupture. Childhood PAN generally has a favorable prognosis compared with adults, though relapses and morbidity are possible. [4, 7] Poor prognostic indicators include persistent proteinuria, renal insufficiency, severe gastrointestinal involvement, cardiomyopathy, and CNS disease. [4,7] Our patient had transient proteinuria without long-term sequelae and remains in remission several months post-treatment.

Conclusion

This case contributes to the limited literature on pediatric PAN by highlighting an unusually young patient with multisystem involvement, biopsy-proven necrotizing vasculitis, and digital gangrene who achieved remission with corticosteroid monotherapy. It underscores the importance of high clinical suspicion and early intervention, particularly in resource-limited settings where angiography may not be feasible. Early

recognition and treatment can significantly improve outcomes and prevent life-threatening complications in cPAN.

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