Clinical Case of Nodular Polyarteritis

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Abstract

Polyarteritis nodosa (PAN) is a rare necrotizing vasculitis affecting small- and medium-sized arteries, leading to vascular inflammation, thrombosis, aneurysm formation, and tissue ischemia. Juvenile-onset PAN is exceptionally rare and often presents with nonspecific manifestations, resulting in delayed diagnosis and irreversible organ damage. We report a case of juvenile-onset PAN with prolonged disease duration complicated by extensive digital necrosis and multiple amputations.

Case presentation: A 42-year-old woman was admitted with necrotic lesions of the cheeks, nose, fingers, and toes, accompanied by cold extremities, dyspnea, chest pain, headache, and generalized weakness. The disease began at the age of 15 years with Raynaud-like symptoms and was managed as Raynaud's syndrome for many years without immunosuppressive therapy. Despite repeated vascular treatment and sympathectomy, progressive ischemia resulted in multiple finger and toe amputations. Laboratory investigations revealed leukocytosis, thrombocytopenia, elevated inflammatory markers, mild renal impairment, and proteinuria. Hepatitis B serology was negative. Cardiac evaluation demonstrated secondary cardiomyopathy, left ventricular hypertrophy, and grade II aortic regurgitation. Based on the characteristic clinical manifestations and multisystem involvement, a diagnosis of juvenile-onset polyarteritis nodosa with moderate disease activity affecting the skin, peripheral vessels, cardiovascular system, kidneys, liver, and joints was established.

Conclusion: This case highlights the diagnostic challenge of juvenile-onset PAN and its potential to mimic Raynaud's syndrome. Persistent digital ischemia, progressive tissue necrosis, and multisystem involvement should raise suspicion for systemic vasculitis. Early diagnosis and timely immunosuppressive therapy are crucial to prevent irreversible vascular damage, amputations, and long-term disability.

Keywords: polyarteritis nodosa; systemic vasculitis; juvenile-onset polyarteritis nodosa; skin necrosis; digital amputation; multisystem involvement; case report.

1. Introduction

Polyarteritis nodosa (PAN), also known as Kussmaul–Maier disease, is a rare systemic necrotizing vasculitis affecting predominantly small- and medium-sized arteries, leading to vascular inflammation, aneurysm formation, thrombosis, and subsequent tissue ischemia. The reported prevalence ranges from 0.7 to 6.3 cases per 100,000 population. Although PAN can occur at any age, severe disease and poor prognosis are associated with early disease onset and male sex; men are affected approximately 2.5 times more frequently than women, with peak incidence between 46 and 50 years of age. First described by Kussmaul and Maier in 1866, PAN primarily involves arterial bifurcations, resulting in microaneurysm formation, vascular occlusion, and infarction of various organs. Long-standing disease may affect multiple systems, including the kidneys, heart, skin, joints, peripheral nervous system, and gastrointestinal tract. The etiology of PAN remains incompletely understood. Associations have been reported with hepatitis B and C viruses, HIV, cytomegalovirus, and parvovirus infections, while certain medications and vaccines have also been implicated as potential triggers. Clinical manifestations are highly heterogeneous and usually begin with nonspecific constitutional symptoms such as fever, weight loss, fatigue, myalgia, and arthralgia, often delaying diagnosis. Organ involvement may present as hypertension and renal dysfunction, coronary vasculitis, peripheral neuropathy, skin lesions including livedo reticularis and necrosis, and gastrointestinal

ischemia. Diagnosis remains challenging because of the absence of a specific laboratory marker. It relies on a combination of clinical findings, laboratory and imaging studies, and histopathological confirmation. Biopsy of affected tissue remains the gold standard, demonstrating segmental necrotizing vasculitis with fibrinoid necrosis and inflammatory infiltration. Current classification is based on the American College of Rheumatology (ACR) criteria and the updated 2022 ACR/EULAR classification criteria. Although PAN predominantly affects adults, juvenile-onset PAN represents a rare subset, with an estimated incidence of less than one case per million children. Compared with adult-onset disease, juvenile PAN more commonly presents with skin manifestations, myalgia, and arthralgia, whereas renal and neurological involvement is less frequent. However, prolonged disease duration may lead to cumulative organ damage, chronic complications, and the need for long-term immunosuppressive therapy.

Clinical case. We report a case of juvenile-onset polyarteritis nodosa (PAN) in a 42-year-old woman who was admitted to the multidisciplinary clinic of Tashkent Medical Academy in 2023. The patient presented with necrotic lesions involving the cheeks, nose, fingers, and toes, coldness of the fingertips, exertional dyspnea, chest pain, palpitations, headache, dizziness, memory impairment, epigastric discomfort, loss of appetite, and general weakness. The disease had begun at the age of 15 years with erythema, cyanosis, cold indurated edema of the feet, lower legs, and palms. Initially, she was diagnosed with Raynaud's syndrome and was followed by vascular surgeons for many years. Despite repeated inpatient and outpatient treatment, her condition progressively worsened. In 2013, a sympathectomy was performed. In July 2022, severe digital ischemia with necrosis of fingers and toes developed, accompanied by intense burning pain and cyanosis of the extremities, requiring multiple amputations of affected digits (Figure 1).

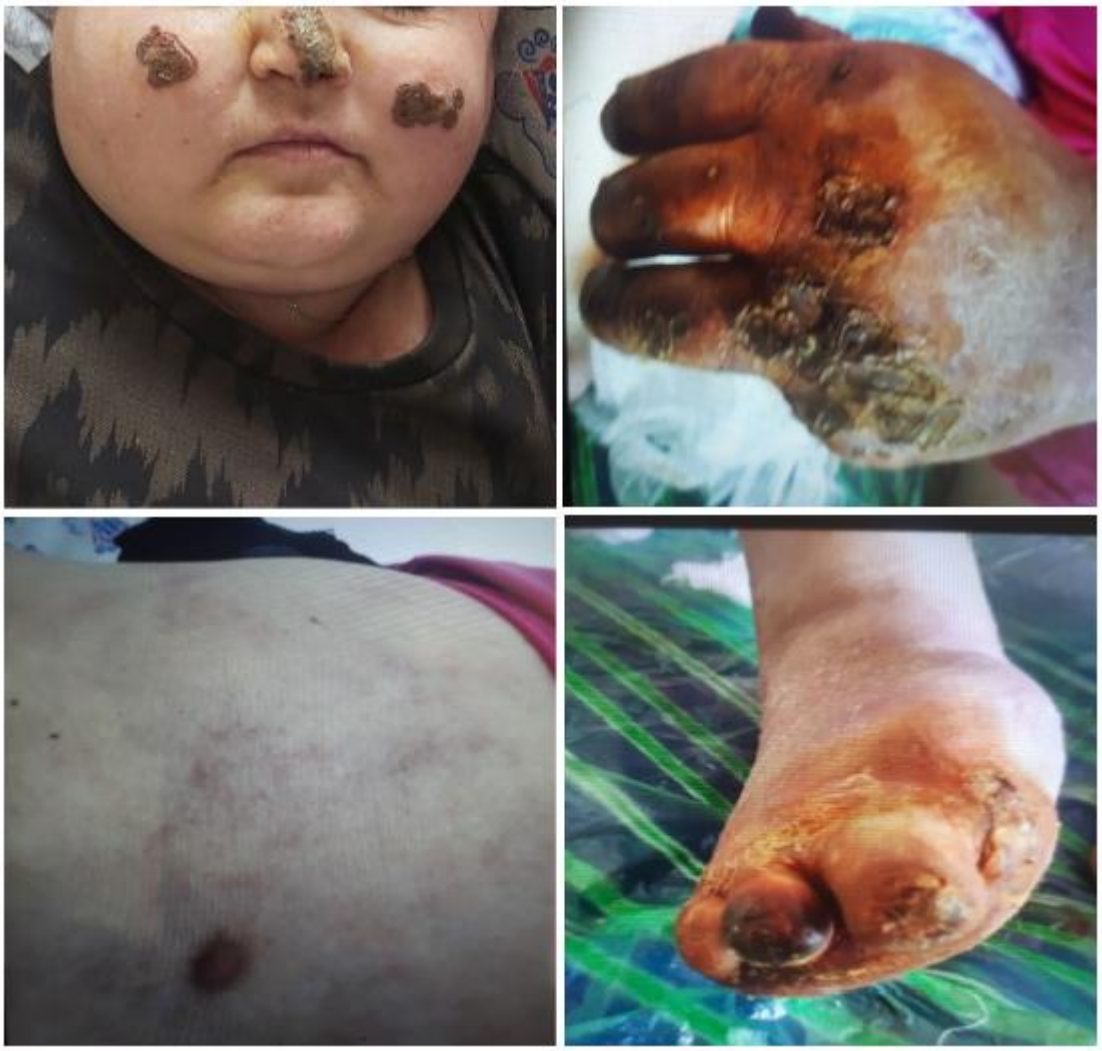

Figure 1. Clinical features (case) of patient with juvenile-onset polyarteritis nodosa.

The patient received antibacterial, vasoactive, and anticoagulant therapy but had never received disease-modifying immunosuppressive treatment. Several months before admission, new necrotic lesions appeared on the cheeks and tip of the nose, prompting hospitalization to the rheumatology department.

Physical examination revealed a patient in moderately severe condition with multiple necrotic lesions on the fingers, toes, cheeks, and nose. Previous amputations included the 2^{nd} – 4^{th} fingers of the right hand, the 4th finger of the left hand, the 3^{rd} – 5^{th} toes of the left foot, and the 3^{rd} and 4^{th} toes of the right foot. No peripheral edema or lymphadenopathy was observed. Laboratory investigations demonstrated leukocytosis (20.5×10^9/L), thrombocytopenia (98×10^9/L), elevated erythrocyte sedimentation rate (40 mm/h), increased C-reactive protein (16 mg/L), mild proteinuria, elevated blood urea (13.4 mmol/L), and borderline elevated serum creatinine (110.7 μmol/L). Hepatitis B serology was negative. Electrocardiography showed sinus bradycardia, left-axis deviation, and repolarization abnormalities. Echocardiography revealed secondary cardiomyopathy, moderate left ventricular hypertrophy, and grade II aortic regurgitation with preserved left ventricular ejection fraction (62%). Ultrasonography demonstrated hepatomegaly with grade I–II hepatic steatosis and renal parenchymal abnormalities. Based on the clinical manifestations, laboratory findings, and multisystem involvement, the patient was diagnosed with juvenile-onset systemic vasculitis consistent with polyarteritis nodosa (activity grade II) involving the skin, cardiovascular system, kidneys, liver, joints, and peripheral blood vessels. The disease course was complicated by extensive ischemic necrosis and multiple digital amputations. The diagnosis was established based on the following diagnostic criteria: weight loss, skin involvement (livedo reticularis), and peripheral neuropathy. The patient received the following treatment regimen: prednisolone at a dose of 0.5–1 mg/kg/day with gradual tapering to a maintenance dose of 5–10 mg/day, in combination with cytostatic agents, including methotrexate 15–25 mg/week and cyclophosphamide administered as monthly intravenous pulse therapy at a dose of 0.5–0.75 g/m². Anticoagulants and antiplatelet agents were also prescribed. As anti-inflammatory therapy for autoimmune inflammation, prednisolone 5 mg was administered orally at a dosage of two tablets three times daily. As baseline immunosuppressive therapy, azathioprine 50 mg was prescribed orally at a dosage of one tablet once daily. Programmed pulse therapy consisted of Allisalon 1000 mL, cyclophosphamide 1000 mg, and 0.9% sodium chloride solution 200 mL, administered intravenously by infusion for a course of three consecutive days. As anticoagulant therapy, Clexane® 0.4 mL was administered subcutaneously for three days.

Conclusion

Polyarteritis nodosa (PAN) remains a significant medical and social challenge, requiring further investigation of its clinical and immunological characteristics, individualized therapeutic approaches, and the development of long-term follow-up strategies. Contemporary treatment modalities, including combinations of glucocorticoids and cytotoxic agents, as well as the use of mycophenolate mofetil, biological agents, and targeted therapies in refractory cases, have substantially improved patient survival. Nevertheless, the long-term outcomes of treatment, quality of life, and issues related to treatment adherence in adult patients with disease onset during childhood remain insufficiently studied.

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