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A Clinicopathological Review of Pediatric Vasculitis: From Cutaneous to Systemic Involvement

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Introduction

Pediatric vasculitis represents a diverse and complex group of disorders characterized by inflammation of blood vessels, leading to tissue ischemia, organ dysfunction and a range of clinical manifestations that differ from those observed in adults. In children, vasculitides encompass both isolated cutaneous conditions and systemic diseases with multi-organ involvement. Understanding the clinicopathological features of pediatric vasculitis is essential for early diagnosis, appropriate management and improved outcomes, especially given the potential for chronic morbidity or life-threatening complications if left untreated or misdiagnosed. This review synthesizes the current understanding of pediatric vasculitis, examining its clinical spectrum, pathological basis, diagnostic challenges and therapeutic strategies across various subtypes ranging from limited skin involvement to systemic syndromes [1].

Description

Cutaneous vasculitis in children most commonly presents as leukocytoclastic vasculitis, characterized by palpable purpura localized to dependent areas such as the lower limbs and buttocks. The majority of these cases are post-infectious or drug-induced and are self-limiting. However, skin involvement may also herald the onset of a systemic vasculitic disorder. Histopathological examination reveals neutrophilic infiltration around post-capillary venules, fibrinoid necrosis of vessel walls and nuclear debris (leukocytoclasia). Direct immunofluorescence may show immune complex deposition, particularly of IgA in Henoch-Schönlein Purpura (HSP), now termed IgA vasculitis. IgA vasculitis is the most common systemic vasculitis in children, typically affecting those aged 3 to 15 years. It is characterized by the classic tetrad of palpable purpura, arthralgia or arthritis, abdominal pain and renal involvement. The underlying pathogenesis involves IgA1-dominant immune complex deposition in small vessels, often triggered by preceding infections, especially upper respiratory tract infections. Renal biopsy in affected children shows mesangial proliferation with IgA deposition, which may mimic IgA nephropathy. While most cases are self-limiting, renal involvement determines prognosis, necessitating careful monitoring and, in severe cases, immunosuppressive therapy [2].

Another important vasculitis in pediatrics is Kawasaki Disease (KD), an acute, self-limiting vasculitis of medium-sized vessels that primarily affects children under five years of age. It is the leading cause of acquired heart disease in children in developed countries. The clinical presentation includes prolonged fever, conjunctival injection, oral mucosal changes (strawberry tongue, cracked lips), extremity changes (erythema and edema), rash and cervical lymphadenopathy. The most feared complication is coronary artery aneurysm formation, which occurs in up to 25% of untreated patients. The pathological hallmark of KD is a panvasculitis of medium-sized arteries with infiltration of neutrophils and mononuclear cells leading to destruction of the internal elastic lamina and medial smooth muscle cells. Timely administration of Intravenous Immunoglobulin (IVIG) and aspirin significantly reduces the risk of coronary involvement. In recent years, Multisystem Inflammatory Syndrome in Children (MIS-C), associated with SARS-CoV-2 infection, has emerged as a novel vasculitic syndrome with features overlapping those of KD, toxic shock syndrome and macrophage activation syndrome. MIS-C presents with persistent fever, gastrointestinal symptoms, rash, conjunctivitis, mucocutaneous inflammation and cardiovascular dysfunction. Laboratory findings often include elevated inflammatory markers, lymphopenia, thrombocytopenia and elevated cardiac enzymes. Cardiac MRI may show myocardial edema or fibrosis and echocardiography may reveal depressed ventricular function or coronary abnormalities. Histopathological data remain limited, but findings of small and medium-vessel vasculitis, perivascular inflammation and endotheliitis have been reported. Immunomodulatory treatment with IVIG, corticosteroids and biologics like anakinra or infliximab has shown efficacy [3,4].

Takayasu Arteritis (TA), although rare in children, constitutes an important cause of large-vessel vasculitis in the pediatric population, especially in adolescents. It predominantly affects the aorta and its main branches and may present insidiously with nonspecific symptoms such as fatigue, weight loss and fever, followed by vascular claudication, pulse deficits, hypertension and bruits. Diagnosis is challenging due to the gradual onset and lack of specific biomarkers. Imaging modalities such as Magnetic Resonance Angiography (MRA) and computed tomography angiography (CTA) are essential for detecting vessel wall thickening, stenosis, or aneurysm formation. Polyarteritis nodosa (PAN) is a necrotizing vasculitis of medium-sized arteries that can affect multiple organs including the skin, gastrointestinal tract, kidneys, peripheral nerves and testes. In children, it may present with fever, myalgia, skin ulcers, hypertension and abdominal pain due to mesenteric ischemia. Cutaneous PAN may be limited to skin and joints but requires differentiation from systemic disease. Histopathology of affected arteries reveals transmural necrotizing inflammation, fibrinoid necrosis and leukocyte infiltration. Pediatric PAN is often associated with hepatitis B virus, although this has become less common due to widespread vaccination. Treatment includes corticosteroids and immunosuppressants like cyclophosphamide in severe cases [5].

Conclusion

In summary, pediatric vasculitis comprises a spectrum of disorders ranging from self-limited cutaneous conditions to severe systemic diseases with multi-organ involvement. Accurate diagnosis relies on a combination of clinical evaluation, laboratory testing, imaging and histopathology. Early recognition and appropriate immunomodulatory therapy are critical for preventing complications and improving long-term outcomes. Ongoing research, including pediatric-specific clinical trials, is needed to optimize diagnostic criteria, treatment protocols and prognostic tools in this complex and evolving field.

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Conflict of Interest

None.

References

1. Petersen, M. Prutz, S. Moller, A. Bygum and A. Voss, et al. "Epidemiology of cutaneous lupus erythematosus and the associated risk of systemic lupus erythematosus: A nationwide cohort study in Denmark." *Lupus* 27 (2018): 1424-1430.
2. Gronhagen, C. M., C. M. Fored, F. Granath and F. Nyberg. "Cutaneous lupus erythematosus and the association with systemic lupus erythematosus: A population-based cohort of 1088 patients in Sweden." *Br J Dermatol* 164 (2011): 1335-1341.
3. Wieczorek, Isabela T., Kathleen J. Propert, Joyce Okawa and Victoria P. Werth. "Systemic symptoms in the progression of cutaneous to systemic lupus erythematosus." *JAMA Dermatol* 150 (2014): 291-296.
4. Gilliam, James N. and Richard D. Sontheimer. "Skin manifestations of SLE." *Clin Rheum Dis* 8 (1982): 207-218.
5. Okon, Lauren G. and Victoria P. Werth. "Cutaneous lupus erythematosus: diagnosis and treatment." *Best Pract Res Clin Rheumatol* 27 (2013): 391-404.

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