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Childhood systemic vasculitis

Erdal Sag [1], Ezgi Deniz Batu [1], Seza Ozen*

Hacettepe University, Ihsan Dogramaci Children's Hospital, Department of Pediatric Rheumatology, Turkey

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A B S T R A C T

Vasculitides are characterized by inflammation of the vessel wall. Most of the vasculitides tend to occur in vessels of a specific size and certain target organs. In this review, we discuss each specific childhood vasculitis according to the latest Chapel Hill Consensus Conference 2012 nomenclature system and the Ankara 2008 classification criteria. We have also reviewed the clinical and laboratory characteristics and the recent treatment recommendations for the vasculitides we encounter in children.

Introduction

Vasculitides are heterogeneous diseases characterized by inflammation of the vessel wall. Primary vasculitides are less common in childhood than in adulthood; the estimated incidence is approximately 50 cases per 100,000 children annually [1]. However, certain types of primary vasculitides occur more often in childhood, such as immunoglobulin A vasculitis/Henoch-Schönlein purpura (IgAV/HSP) and Kawasaki disease (KD) [2]. See Fig. 1.

Nomenclature system and classification of systemic vasculitides

Because primary vasculitides have a heterogeneous nature, it is important but difficult to classify them. A nomenclature system is essential to provide names and definitions and form a framework for the diagnostic/classification criteria [3]. In 1994, an international conference was held in Chapel Hill (Chapel Hill Consensus Conference [CHCC]) to develop a nomenclature system for vasculitides [4]. In

* Corresponding author. Hacettepe University, Ihsan Dogramaci Children's Hospital, Department of Pediatric Rheumatology, 06230, Sihhiye, Ankara, Turkey.
E-mail address: sezaozen@hacettepe.edu.tr (S. Ozen).
[1] The first two authors contributed equally to this work.

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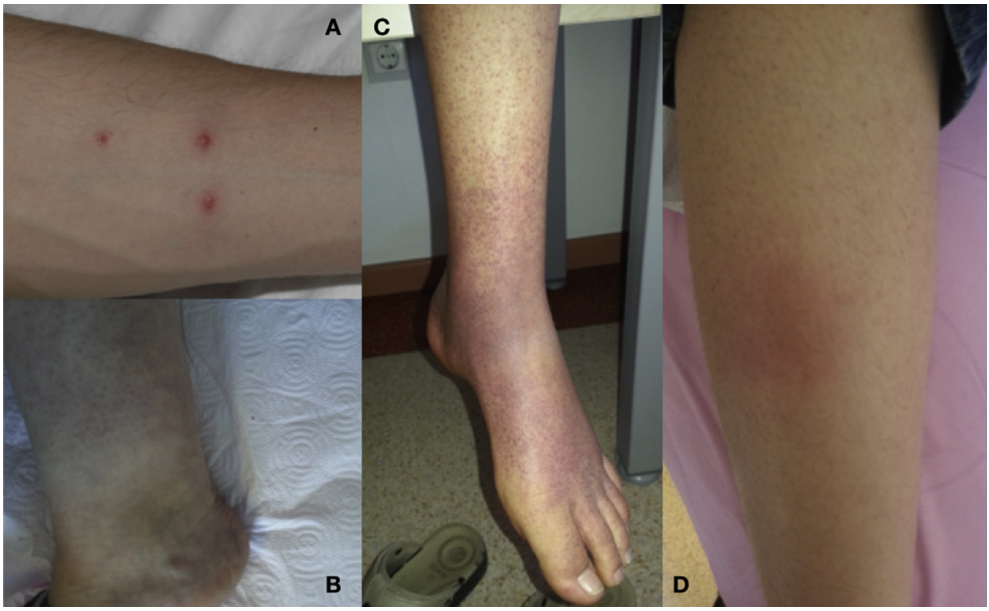

Fig. 1. Cutaneous features of childhood vasculitis. **A**. A positive pathergy test in Behçet's disease, **B.** Livedo reticularis in a PAN patient, **C.** Palpable purpura in IgAV/HSP, **D**. Erythema nodosum in Behçet's disease.

the CHCC system, vasculitides were categorized according to the size of the vessels predominantly affected. In 2012, this nomenclature was updated by replacing most of the eponyms with the terms that describe the disease pathogenesis and by adding some new categories such as variable vessel vasculitis (VVV), single-organ vasculitis, vasculitis associated with systemic disease, and vasculitis associated with probable etiology (Table 1) [5].

In primary vasculitis, we usually do not have single pathognomonic tests to diagnose our patients; thus, we have classification criteria but not diagnostic criteria for most of them. Diagnostic criteria are applied to the individual patient to diagnose the disease by ruling out other mimicking conditions [6], while classification criteria are mainly for defining more homogeneous patient groups for research purposes [7]. The American College of Rheumatology (ACR) had proposed a series of classification criteria for vasculitis in adults in 1990 [8,9]. However, the ACR criteria set has never been validated in children [8–13]. The classification criteria for the most common vasculitides in childhood such as IgAV/HSP, KD, polyarteritis nodosa (PAN), granulomatous polyangiitis/Wegener granulomatosis (GPA/WG), and Takayasu arteritis (TA) were developed and validated at the 2008 Ankara Consensus Conference by the endorsement of European League Against Rheumatism (EULAR), Pediatric Rheumatology European Society, and the Pediatric Rheumatology International Trials [14–16]. The main characteristics of the ACR [10–13] and Ankara 2008 classification criteria [15] are summarized in Table 2.

Currently, there are no diagnostic criteria available for most of the primary systemic vasculitis [17]. The "diagnostic and classification criteria for vasculitides (DCVAS)" study in adult patients is currently underway to develop and validate diagnostic criteria and improve classification criteria [18].

I Large-vessel vasculitis

Large-vessel vasculitis (LVV) is defined as vasculitis that predominantly affects large arteries [5]. LVV includes giant cell arteritis (GCA) and TA [5].

Ia Giant cell arteritis.

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