Gastrointestinal and Hepatic Disease in Vasculitis

Eric Anderson, MD, Neel Gakhar, MD, Courtney Stull, MD, Liron Caplan, MD, PhD*

KEYWORDS

• Vasculitis • Granulomatosis with polyangiitis • Polyarteritis nodosa
• Antineutrophil cytoplasmic antibody–associated vasculitis • Kawasaki disease
• Gastrointestinal diseases • Mouth diseases • Esophageal diseases

KEY POINTS

- Vasculitis can affect every organ of the gastrointestinal system.
- Vasculitis often presents with nonspecific symptoms, including gastrointestinal bleeding, and should be included in the differential when initial work-up fails to find an obvious source of gastrointestinal symptoms.
- Gastrointestinal involvement in the setting of systemic vasculitis often represents more severe disease making early diagnosis and treatment imperative.

INTRODUCTION

The vasculitides are a group of disorders that cause inflammation of blood vessels. They can be classified according to the size of the arteries affected (small, medium, and large), etiology (primary vs secondary), and extent of involvement (localized vs

Conflicts of Interest: All authors declare no conflicts of interest in this article, including financial, consultant, institutional, and other relationships that might lead to bias or a conflict of interest.
Guarantor of the Article: Liron Caplan, MD, PhD.
Financial Support: There was no financial or grant support for this study. Dr L. Caplan is supported by VA HSR&D IIR 14-048-3 and the Michelson Fund at the University of Colorado Foundation.
Author Contributions: E. Anderson, N. Gakhar, C. Stull, and L. Caplan were responsible for drafting, critical revision, and approval of the final article. All authors were involved in the final approval of the version of the article submitted and have agreed to be accountable for all aspects of the work. The views expressed in this article are those of the authors and do not necessarily reflect the position or policy of the Department of Veterans Affairs.
Potential Competing Interests: None.
Denver Veterans Affairs Medical Center, University of Colorado School of Medicine, Denver, CO, USA
* Corresponding author. Denver Veterans Affairs Medical Center, University of Colorado School of Medicine, 1775 Aurora Court, B115, Aurora, CO 80045.
E-mail address: liron.caplan@ucdenver.edu

Rheum Dis Clin N Am 44 (2018) 1–14
https://doi.org/10.1016/j.rdc.2017.09.001
0889-857X/18/Published by Elsevier Inc.

systemic). If untreated, this group of diseases can potentially cause life-threatening complications with significant morbidity. Therefore, it is vital that clinicians identify and treat this group of diseases effectively. Unfortunately, the scientific basis for summarizing the digestive and hepatic involvement by vasculitides is largely limited to case reports and case series.

This article examines the different types of vasculitis that may affect each anatomic component of the gastrointestinal system. The initial section summarizes the nomenclature, classification, and diagnostic criteria for some of the vasculitides. The epidemiology of vasculitis is addressed and then individual sections are dedicated to anatomic components of the gastrointestinal system, touching on the particular types of vasculitis known to affect each gastrointestinal organ. These sections address the presentation, pathophysiology, and treatment of vasculitis affecting each organ system.

If affected by vasculitis, the oral mucosa and esophagus are typically involved with Kawasaki disease (KD) or the antineutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAVs). These include granulomatosis with polyangiitis (GPA), eosinophilic GPA (EGPA), and microscopic polyangiitis (MPA). The stomach may also be affected by the AAV and by leukocytoclastic vasculitis (LCV), in particular, immunoglobulin A–associated vasculitis (IgAV), formerly known as Henoch-Schönlein purpura. The small intestine, large intestine, and mesentery can be affected by Takayasu arteritis (TA), LCV, AAV, polyarteritis nodosa (PAN), giant cell arteritis (GCA), systemic lupus erythematosus (SLE)-associated vasculitis, and rheumatoid arthritis–associated vasculitis (RAAV). The liver and gall bladder are typically affected by PAN and single-organ vasculitis (SOV), but there are numerous reports of involvement from other vasculitides as well, including GCA. Finally, the pancreas can also be involved with GPA and SOV. Behcet's disease will not be addressed here, as there is a dedicated article to this vasculitis and its gastrointestinal manifestations (see Ibrahim Hatemi and colleagues' article, "Gastrointestinal Involvement in Behçet Disease," in this issue). SLE-associated vasculitis is only briefly mentioned, for the same reason.

NOMENCLATURE, CLASSIFICATION CRITERIA, AND DIAGNOSTIC CRITERIA

GPA represents a necrotizing vasculitis typically affecting small and medium-sized vessels. It commonly involves the respiratory tract and kidneys, where it causes a necrotizing glomerulonephritis. In 1990, the American College of Rheumatology (ACR) established classification criteria for Wegener granulomatosis, the eponym previously assigned to GPA.[1] These criteria include the following:

- Nasal or oral inflammation
- Abnormal chest radiograph with infiltrates or nodules
- Urinary sediment with microhematuria (greater than 5 white blood cells per high-power field)
- Granulomatous inflammation on biopsy of artery or arteriole

Two or more of these criteria confer an 88.2% sensitivity and 92% specificity for establishing a diagnosis of GPA.[2]

EGPA is also a necrotizing vasculitis that affects small and medium-sized vessels, which is associated with asthma and eosinophilia. The ACR established 6 criteria to classify EGPA and they are as follows:

- Asthma
- Eosinophilia (more than 10% of circulating leukocytes)
- Paranasal sinusitis
- Pulmonary infiltrates on radiographs