Medical Sciences

Churg-Strauss Syndrome Mimicking Parasitic Infestation: Atypical Presentation of A Rare Disease

Patompong Ungprasert[1,2], Wisit Cheungpasitporn[3,*], Charat Thongprayoon[3], and Narat Srivali[3]

[1]Department of Medicine, Bassett medical center, Cooperstown, NY, USA. [2]Department of medicine, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand. [3]Department of Medicine, Mayo Clinic, Rochester, MN, USA.

Context: Churg-Strauss syndrome (CSS) is a rare antineutrophil cytoplasmic antibody associated vasculitis characterized by asthma, chronic rhinosinusitis, and persistent eosinophilia. Although acute diarrhea is frequent clinical manifestations of parasitic infection, it is not a main clinical presentation of CSS. We report a 49-year-old man who presented with acute watery diarrhea and peripheral eosinophilia. Extensive investigations for parasitic infestation were negative. He subsequently underwent colonoscopy which showed numerous serpiginous ulcers throughout the colon. Microscopic examination revealed ulcerated colonic mucosa with eosinophilic infiltration. Further serological investigation demonstrated a positive myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA). The patient was diagnosed with CSS and treatment with high dose oral corticosteroid was initiated and gradually tapered. His diarrhea, shortness of breath, rash, eosinophilia and pulmonary infiltration dramatically improved after the treatment. Conclusion: Although gastrointestinal symptoms are not main clinical presentations of CSS. Acute diarrhea with peripheral eosinophilia should raise the suspicious of CSS, especially after excluding the possibility of parasitic infestation. *Journal of Nature and Science, 1(5):e105, 2015*

ANCA vasculitits | Churg-Strauss Syndrome | Parasitic infection | Vasculitis

Introduction

Churg-Strauss syndrome (CSS), also known as eosinophilic granulomatosis, is a rare antineutrophil cytoplasmic antibody associated vasculitis characterized by asthma, chronic rhinosinusitis, and persistent eosinophilia [1]. It is typically described as 3-stage disease with the prodromal phase of asthma, the second phase of peripheral blood and tissue eosinophilia and the third phase of vasculitis. Diagnosis of CSS can be challenging and sometimes difficult to distinguish from hypereosinophilic syndrome (HES) as it can mimic the clinical manifestations of CSS, including blood eosinophilia, peripheral neuropathy and eosinophilic infiltration in various organs. However, asthma and atopic symptoms are extremely uncommon in patients with HES as none of these patients were found to have asthma in 2 large cohort studies though few sporadic cases of asthma in patients with HES have been reported [2].

Gastrointestinal (GI) symptoms such as diarrhea and abdominal pain are not main clinical presentations of CSS and American College of Rheumatology (ACR) has not included GI manifestations in their criteria for diagnosis of CSS [2-4]. Conversely, acute diarrhea with peripheral eosinophilia are frequent clinical manifestations of parasitic infection such as *Strongyloides stercoralis* [5]. We report a challenging case of CSS who presented with acute watery diarrhea.

Case Report

A 49-year-old man presented to our institute with a 5-day history of profuse watery diarrhea without any associated abdominal pain or fever. His past medical history was significant for adult-onset asthma that was poorly controlled by inhaled corticosteroid. He also had a frequent travel history to Mexico. His last trip was 3 months prior to the onset of symptoms. Physical examination was remarkable for wheezing in both lungs. Initial laboratory investigations were remarkable for a marked peripheral eosinophilia of 6570 cells/uL (reference range, 5-500 cells/uL), though there was no previous complete blood cell count to compare, and an elevated ESR of 67 mm/hr. Chest x-ray revealed bilateral scattered ground glass opacities with mild bilateral hilar adenopathy. Other blood chemistry tests, urinalysis, electrocardiogram and echocardiogram were unremarkable. Parasitic infestation, particularly *Strongyloides stercoralis* [5], was strongly suspected. However, extensive investigations for parasitic infestation, including direct microscopic exam for ova and parasite, stool giardia and cryptosporidium antigen, strongyloides IgG and IgM, and stool culture, were all negative. He subsequently underwent colonoscopy which showed numerous serpiginous ulcers throughout the colon (Figure 1). Microscopic examination revealed ulcerated colonic mucosa with eosinophilic infiltration (Figure 2). On the fourth day of admission, he developed purpuric rash over his shin, which was biopsied and was shown to be a leukocytoclastic vasculitis [6, 7]. Further serological investigation demonstrated a positive myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA). The patient was finally diagnosed with Churg-Strauss syndrome and treatment with high dose oral corticosteroid was initiated and gradually tapered. His diarrhea, shortness of breath, rash, eosinophilia and pulmonary infiltration dramatically improved after the treatment.

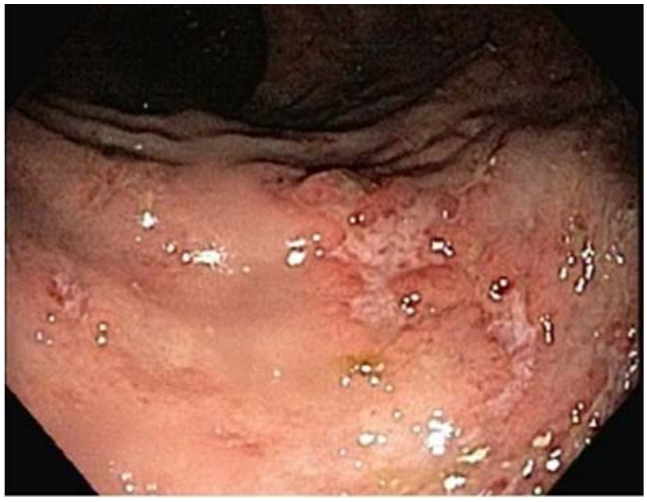

Figure 1: Colonoscopy revealed numerous serpiginous shallow ulcers throughout the colon.

Discussion

In this case, the diagnosis of CSS was confirmed by colonoscopy as the eosinophilic infiltration seen on the microscopic examination of the biopsied colonic mucosa, along with the presence of asthma, peripheral eosinophilia and pulmonary infiltration, fulfilled 4 out of 6 ACR criteria for the diagnosis of CSS [2]. The patient's adult-onset asthma and positive MPO-ANCA additionally helped to distinguish CSS from HES.

Conflict of interest: No conflicts declared.
* Corresponding Author. Wisit Cheungpasitporn, MD. Address: 200 First Street SW, Mayo Clinic, Rochester, MN, USA. 55905.
E-mail: wcheungpasitporn@gmail.com

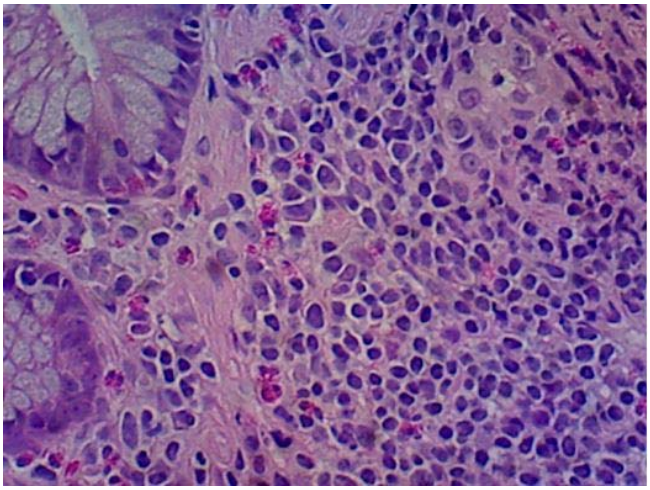

Figure 2: Microscopic examination of the ulcer demonstrated eosinophilic infiltration in the colonic mucosa (hematoxylin-eosin)

Gastrointestinal symptoms, though not a prominent feature or a direct ACR criterion [3, 4], are also common in patients with this syndrome with a reported incidence of 33% [2,8]. In a recent case series by Pagnoux *et al.*[9], abdominal pain was the most common symptom which was seen in more than 90% of patients with CSS who had gastrointestinal tract involvement whereas diarrhea and gastrointestinal bleeding were seen in half of patients.[9] Nevertheless, colonic ulcer, which was seen in this case, is infrequently described with fewer than 20 reported cases in the literatures [9]. These ulcers are believed to be a consequence of bowel ischemia from mesenteric artery vasculitis though eosinophilic cationic protein released by the infiltrating eosinophils might also directly insult the gastrointestinal mucosa. Interestingly, for unknown reason, most of the previous reported cases are from Japan[10] and, to the best of our knowledge, this is the second reported case of CSS presenting with colonic ulcer in the United States [11]. CSS with recurrent small bowel obstruction and choledocholithiasis was also recently reported; therefore physicians should be aware of GI manifestations of CSS that may allow for timely management of this disorder [12].

Corticosteroid remains the cornerstone of the treatment of CSS. However, if the patient has a high risk feature as indicated by having at least two out of the five factors score [13] (cardiac involvement, gastrointestinal involvement, renal insufficiency, proteinuria, and central nervous system involvement) or having the five factor score of one with cardiac or central nervous system involvement, cyclophosphamide is preferred [11]. In this patient, with the five factor score of only one (colonic involvement), oral corticosteroid was utilized with a favorable clinical outcome.

In summary, our case also underscores the importance of CSS as a potential cause of diarrhea with eosinophilia after infectious etiology is excluded. A detailed history, particularly history of adult-onset asthma, and thorough physical examination can provide a pivotal clue to the timely diagnosis and prompt treatment for this relatively uncommon syndrome.

Authors' contributions

All authors had access to the data and a role in writing the manuscript.

1. Cartin-Ceba R, Keogh KA, Specks U, Sethi S, Fervenza FC: Rituximab for the treatment of Churg-Strauss syndrome with renal involvement. *Nephrol Dial Transplant* 2011, 26(9):2865-2871.
2. Ungprasert P, Srivali N, Cheungpasitporn W, Schaeffer CS: Is it acute coronary syndrome or Churg-Strauss syndrome? *Am J Emerg Med* 2013, 31(1):270 e275-278.
3. Lanham JG, Elkon KB, Pusey CD, Hughes GR: Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg-Strauss syndrome. *Medicine (Baltimore)* 1984, 63(2):65-81.
4. Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP, Calabrese LH, Edworthy SM, Fauci AS, Leavitt RY *et al*: The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990, 33(8):1094-1100.
5. Ahmed S, Rashid S, Ammannagari N, Cheungpasitporn W: Chasing Eosinophilia in Loeffler's Syndrome: A Case of Strongyloidiasis in Upstate New York. *N Am J Med Sci* 2013, 5(3):248.
6. Cheungpasitporn W, Jirajariyavej T, Howarth CB, Rosen RM: Henoch-Schonlein purpura in an older man presenting as rectal bleeding and IgA mesangioproliferative glomerulonephritis: a case report. *J Med Case Rep* 2011, 5:364.
7. Srivali N, Ungprasert P, Ahmed S, Cheungpasitporn W, Bischof EF: A case of childhood vasculitis presenting in adulthood. *Am J Emerg Med* 2013, 31(1):254-255.
8. Ungprasert P CS, Kittanamongkolchai W, Cheungpasitporn W: In-Depth Review of ANCA-associated Vasculitis. In: *Advances in Medicine and Biology. Volume 68*, edn. Edited by Berhardt LV: Nova Science Publishers; 2013: 159-172.
9. Pagnoux C, Mahr A, Cohen P, Guillevin L: Presentation and outcome of gastrointestinal involvement in systemic necrotizing vasculitides: analysis of 62 patients with polyarteritis nodosa, microscopic polyangiitis, Wegener granulomatosis, Churg-Strauss syndrome, or rheumatoid arthritis-associated vasculitis. *Medicine (Baltimore)* 2005, 84(2):115-128.
10. Memain N, De BM, Guillevin L, Wechsler B, Meyer O: Delayed relapse of Churg-Strauss syndrome manifesting as colon ulcers with mucosal granulomas: 3 cases. *J Rheumatol* 2002, 29(2):388-391.
11. Berarducci M, Thomas C, Kay J: Churg-strauss syndrome with diffuse gastrointestinal involvement. *J Clin Rheumatol* 1996, 2(4):221-226.
12. Franco DL, Ruff K, Mertz L, Lam-Himlin DM, Heigh R: Eosinophilic granulomatosis with polyangiitis and diffuse gastrointestinal involvement. *Case reports in gastroenterology* 2014, 8(3):329-336.
13. Guillevin L, Lhote F, Gayraud M, Cohen P, Jarrousse B, Lortholary O, Thibult N, Casassus P: Prognostic factors in polyarteritis nodosa and Churg-Strauss syndrome. A prospective study in 342 patients. *Medicine (Baltimore)* 1996, 75(1):17-28.