Case Report

DOI: 10.14235/bas.galenos.2018.2716
Bezmialem Science 2019;7(3):255-8

Granulomatosis Polyangiitis Case that Mimics Henoch-Schönlein Purpura

Tahsin KARAASLAN, Cumali KARATOPRAK

Bezmialem Vakıf University Faculty of Medicine, Department of General Internal Medicine, İstanbul, Turkey

ABSTRACT

Granulomatosis polyangiitis (GPA) is a systemic, necrotizing, granulomatous, antineutrophil cytoplasmic antibody (ANCA) -associated vasculitis that affects small and medium arteries, mainly affecting the upper and lower respiratory tract and the kidneys. It is usually seen over 40 years old. Diagnosis is based on clinical findings, cytoplasmic- C-ANCA positivity and histological findings. Here we report a case of 21-year-old patient who presented with petechial purpuric lesions, abdominal pain, large joint arthritis and hematuria- proteinuria and to whom we started treatment for Henoch-Shönlein Purpura. But a chest imaging showed mass lesion and our final diagnosis was atypical GPA after excluding malignancy, cryoglobulinemia and ANCA related vasculitis in differantial diagnosis.

Keywords: Henoch-Schönlein purpura, atypical granulomatous polyangiitis, ANCA associated vasculitis

Introduction

Granulomatous polyangiitis (GPA), formerly known as Wegener granulomatosis, is an antinutrophil cytoplasmic antibody (ANCA) associated systemic necrotizing granulomatous vasculitis that involves small and medium diameter vessels, mainly affecting the upper and lower respiratory tract and kidneys (1). It has 2 types: Limited GPA and diffuse GPA. Limited GPA is the type of vasculitis that involves upper or lower respiratory tract or involves only eye (2). Proteinase-3 enzyme is the target antigen of C-ANCA and C-ANCA is considered very sensitive. C-ANCA is 90% positive in the active period and this rate decreases in remission (3). Henoch-Schonlein purpura (HSP) is a systemic leukocytoclastic vasculitis characterized by the storage of immunoglobulin A containing immune complexes and complement components in small vessel walls. Although skin, joint, gastrointestinal tract and kidney involvement are at the forefront, other organs such as brain, lung and scrotum can be involved during the course of HSP (4). We wanted to present a case presenting with palpable purpura, severe abdominal pain, arthritis in large joints and hematuria-proteinuria suggesting HSP, but who was diagnosed as having GPA based on the involvement pattern, kidney biopsy findings and C-ANCA positivity.

Case Report

A 21-year-old female patient admitted to our emergency service with throat sore, difficulty in swallowing, cough, bloody phlegm, abdominal pain, common joint pains, rash in the buttocks and legs starting three weeks ago. In physicial examination; fever was 39 °C, blood pressure 110/70 mmHg, heart rate 124/min. She had pale skin, 3x1 cm aphthous ulcer on the back wall of the pharynx, 2 cm splenomegaly on the costa broadcast and palpable purpura on both lower extremities. Perianal ulcers were present (Figure 1). She had proteinuria (+++) and hematuria in urine examination. Other laboratory data were given in Table 1. The amount of protein in 24-hour urine was 1380 mg. Neck computed tomography (CT) showed 23x13 mm lymphadenomegalies (LAM) in all zones, of which the largest ones were in posterior cervical triangle and the carotid space. Thorax CT showed consolidated areas in all zones of bilateral lungs which were diffuse, were in icy glass density, tended to merge and were suggestive of

Address for Correspondence: Tahsin KARAASLAN, Bezmialem Vakıf University Faculty of Medicine, Department of General Internal Medicine, İstanbul, Turkey
Phone: +90 505 935 11 22 **E-mail:** drtkaraaslan@hotmail.com **ORCID ID:** orcid.org/0000-0002-1529-1790

Received: 28.08.2018
Accepted: 03.11.2018

Cite this article as: Karaaslan T, Karatoprak C. Granulomatosis Polyangiitis Case that Mimics Henoch-Schönlein Purpura. Bezmialem Science 2019;7(3):255-8.

alveolitis. In the upper right lung, a 30x22 mm irregular soft tissue mass was observed and a 13x12 mm irregular bordered satellite nodule was observed near it (Figure 2). Diffuse wall thickening was observed in caecum, ascending colon, hepatic flexura, transverse and descending colons in abdominal CT. In the right iliac chain, heterogeneous contrasting soft tissue lesion approximately 30x20x48 mm in size was observed. Mesenteric LAMs in size of 17x12 mm were observed. Acid-resistant bacillus was found negative in the phlegm and no reproduction was observed in haemocultures. The Quantiferon test was negative. Immune globulins were detected in normal range. The result of cryoglobulin was negative. The skin biopsy showed intraepidermal pustular formation and leukocytoclastic vasculitis. Ig, complement, and fibrinogen accumulation were not observed by immunofluorescence method. P-ANCA was negative, whereas C-ANCA was positive (143 pg/mL). The kidney biopsy showed a crescentic glomerulonephritis (GN) without accumulation of immune complexes which was compatible with the diagnosis of GPA. For three days, 500 mg pulsed steroid and endoxan were given. The patient's steroid treatment was continued to be 1 mg/kg after pulse steroid treatment. After three days of treatment, the patient's oral ulcers disappeared and she began to eat, the rash of the patient improved, the findings of arthritis were lost and the patient was transferred to rheumatology and nephrology departments for follow-up and treatment.

Figure 1. Patient's rash screen

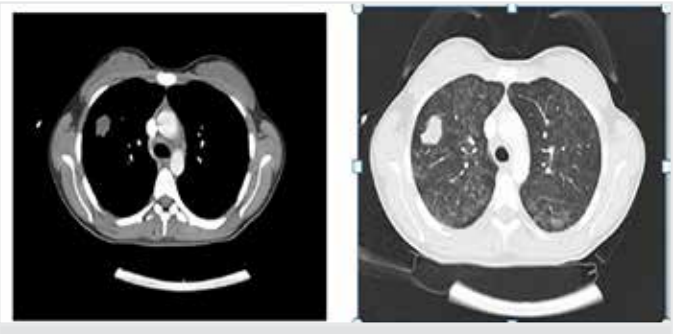

Figure 2. Tharax computed tomography image

Discussion

Vasculitis is a group of inflammatory diseases which have different clinical and pathological characteristics and lead to organ failure with inflammation, necrosis and damage to the walls of the vessels of various sizes and types. They can be fatal without early diagnosis and proper treatment. Immune deposits on the vessel wall are minimal or absent in the ANCA- associated vasculitis (pauci-immune vasculitis). ANCA-related vasculitis is classified as granomatous polyangiitis (Wegener's), eosinophilic granulomatous polyangiitis (Churg-Strauss), microscopic polyangiitis and one-organ limited vasculitis (for example renal limited vasculitis) (5). Lung involvement may be asymptomatic or acute and fulminant. It may present with alveolar haemorrhage causing respiratory insufficiency. Nodules, cavity formations, and infiltrations can be seen frequently in the lung X-ray or in the high-resolution computed tomography scan of the lungs (6). Fever, fatigue, sweating, weight loss (>10%), myalgia, arthralgia are general complaints. About half of the cases have vasculitic skin lesions. The most frequently observed skin symptom is palpable purpura, which shows pathology of leukocytoclastic vasculitis (7). Arthralgia, seen in approximately 70% of patients, usually affects the major joints of lower extremity. Arthritis is less common (8). GPA's gastro-intestinal (GI) involvement may be asymptomatic, as well as gastroenteritis, GI bleeding, ulcer, perforation, cholecystitis, acid, pancreatitis, pancreatic mass and perianal ulcer can be seen (9). In the literature, we found two cases with GPA who presented similar to our case with HSP (10). We know that ANCA is positive in 90% of patients with systemic GPA and 60% of patients with limited GPA. However, it is important to remember that ANCA positivity is not only seen in ANCA-associated vasculitis, but can also be seen in many other diseases and during drug treatments. Biopsy negativity or ANCA positivity does not always exclude the diagnosis of GPA. Appropriate biopsy sites for diagnosis of GPA are upper respiratory tract (especially sinuses), skin, lung and kidneys. Due to the insufficiency of wegener diagnostic criteria of the American College of Rheumatology in 1990, the following criteria were defined at the Chapel Hill Consensus Conference in 1992.

- Granulomatous inflammation of the respiratory tract
- Vasculitis involving small and medium diameter vessels,
- Necrotizing GN
- C-ANCA positivity

In our 21-year-old case; abdominal pain, iron deficiency anemia with fecal occult blood test positivity, microscopic hematuria, non-thrombocytopenic palpable purpura, arthritis and arthralgia suggested the diagnosis of HSP. Alveolar hemorrhage can be seen in HSP, but nodules and formation of cavity are not common in HSP. We evaluated ANCA-associated vasculits, tuberculosis, lymphoma, sarcoidosis, cryoglobulinemic vasculitis and viral infections in differential diagnosis due to oral ulcers, alveolar consolidation and nodules in the lungs in Thorax CT and widespread LAMs in the patient. The skin biopsy showed leukocytoclastic vasculitis, which helped us narrow the differential diagnosis.

Table 1. Laboratory findings of the patient

	28.04.2018	29.04.2018	1.05.2018	Normal range
Glucose	99		89	70-105 mg/dL
BUN	21		20	7-18 mg/dL
Creatinin	0.8		0.67	0.57-1.11 mg/dL
AST	26		26	5-34 U/L
ALT	28		62	0-55 U/L
LDH	247		355	125-220 U/L
Total protein		5.6	4.9	6.2-8.1 g/dL
Albumin		3.1	2.7	3.5-5 g/dL
Hb	7.3		7.4	12.2-16.2
Hct	23		23.9	35.5-48
MCV	70		72	80-97
WBC	14.100		13.100	4.6-10.2
PLT	495.000		369.000	142-424
Urinary analysis	Protein (+++), 34 erytrocytes			
CRP		11	11,9	<0.5 mg/dL
Procalsitonin		2.12	4.82	<0.5 ng/mL
Na	135		139	mmol/L
K	4.8		4.5	mmol/L
HBsAg	Negative			Negative
Anti-HBs	Negative			Negative
Anti-HCV	Negative			Negative
Anti-HIV	Negative			Negative
ANA	Negative			Negative
Anti-ds DNA	Negative			Negative
Anti ENA scl70	Negative			Negative
Anti Sm	Negative			Negative
Antiphospholipid	Negative			Negative
Anti-cardiolipin IgM	Negative			Negative
Anti-cardiolipin IgG	Negative			Negative
ANCA	Positive, 1:100			Negative <1:10
p-ANCA	Negative			Negative
c-ANCA	Positive, 161			Negative, <12 IU/mL
Complemant C3	107			89-193 mg/dL
Complemant C4	15			15-57 mg/dL
GBMA	Negative			
LA	Negative			
Quantiferon	Negative			
IgA	56			65-421 mg/dL
IgM	873			552-1631 mg/dL
IgG	32			33-293 mg/dL
Protein in 24-hour urine	1306		2416	-300 mg/gün

BUN: Blood urea nitrogen, AST: Aspartate aminotransferase, ALT: Alanin aminotransferaz, LDH: Laktat dehidrogenaz, MCV: Mean cell volume, WBC: White blood cell, PLT: Platelet, CRP: C-reactive protein, ANA: Nurses association, HIV: Human immmunodeficiency virus, IgM: Immunoglobulin M, IgG: Immunoglobulin G, GBMA: Generic Biosimilar Medicines Association, Hb: Hemoglobin, ANCA: Antinutrophil cytoplasmic antibody, IgA: Immunoglobulin A

C-ANCA positivity supported the diagnosis of GPA. Significant mucosal thickening of the sinuses in the paranasal sinus x-ray and large oral ulcers suggested the diagnosis of GPA. Urine analysis findings showed renal involvement. The kidney biopsy showed a crescentic GN without accumulation immune complexes which suggested the diagnosis of GPA. Perianal ulcers were also compatible with GPA as reported in the literature.

As a result, we could start treatment for HSP with the existing diagnostic criteria in our case with abdominal pain, rashes, joint involvement and appropriate age suggesting HSP. However, as in our case, taking into account several incompatible findings, we achieved a diagnosis that is more aggressive and can result in death if not treated quickly as a result of extensive research. We wanted to emphasize that the first diagnosis could be misleading and that even the smallest hint should be investigated.

Ethics

Informed Consent: A consent form was completed by all participants.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: T.K., C.K., Design: T.K., C.K., Data Collection or Processing: T.K., Analysis or Interpretation: T.K., Literature Search: T.K., Writing: T.K.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. LutaloPM, D'Cruz DP. Diagnosis and classification of granulomatosis with polyangiitis (aka Wegener's granulomatosis). J Autoimmun 2014;48-49:94-8.
2. Holle JU, Gross WL, Holl-Ulrich K, Ambrosch P, Noelle B, Both M, et al. Prospective long-term follow-up of patients with localised Wegener's granulomatosis: does it occur as persistent disease stage? Ann Rheum Dis 2010;69:1934-9.
3. Csernok E. Antineutrophil cytoplasmic antibodies and pathogenesis of small vessel vasculitides. Autoimmun Rev 2003;2:158-164.
4. Koçak M, Büyükkaragöz B, Kuraş Can Y, Çelebi Tayfur A, Çaltık Yılmaz A, Günbey S. The Epidemiological, Clinical and Laboratory Features of 91 Children with Henoch-Schönlein Purpura. Abant Medical Journal 2015;4:134-40.
5. J. C. Jennette, R. J. Falk, P. A. Bacon, N. Basu, M. C. Cid, F. Ferrario ,et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum 2013;65:1-11.
6. Martinez F, Chung JH, Digumarthy SR, Kanne JP, Abbott GF, Shepard JA et al. Common and Uncommon Manifestations of Wegener Granulomatosis at Chest CT: Radiologic-Pathologic Correlation. RadioGraphics 2011; 32;51-69.
7. Daoud MS1, Gibson LE, DeRemee RA, Specks U, el-Azhary RA, Su WP. Cutaneous Wegener's granulomatosis: clinical, histopathologic, and immunopathologic features of thirty patients. J Am Acad Dermatol 1994;31:605-12.
8. Jeleniewicz R, Suszek D, Majdan M. Musculoskeletal symptoms in a group of granulomatosis with polyangiitispatients. Wiad Lek 2018;71:17-20.
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;84:115-28.
10. Bui T, Chandrakasan S, Poulik J, Fathalla BM. Granulomatosis with polyangiitis presenting as Henoch-Schönlein purpura in children. J Clin Rheumatol 2013;19:199-202.