

Microscopic polyangiitis complicated with bilateral brachial plexopathy: a case report and review of the literature

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Abstract Microscopic polyangiitis is a small-vessel necrotizing vasculitis associated with anti-neutrophil cytoplasmic antibodies and presents itself with glomerulonephritis and hemorrhagic pulmonary capillaritis. Peripheral nervous system involvement is common in anti-neutrophil cytoplasmic antibodies-associated vasculitis, but brachial plexopathy is unusual. We present the case of a 22-year-old man with known microscopic polyangiitis who was under maintenance therapy with prednisolone and cyclophosphamide and developed cough, dyspnea, and hemoptysis which increased in 6 days accompanying pain and paresthesia in the upper limbs. His physical examination revealed hypoesthesia, absence of deep tendon reflexes, and decreased muscle strength in the upper limbs. His chest computed tomography scan showed ground glass pattern in the lower and middle lobes. Electromyography and nerve conduction study showed bilateral brachial plexopathy with involvement of all of the cervical roots that were more severe in the lower roots and left side.

Keywords Microscopic polyangiitis · Vasculitis · Brachial plexopathy

Introduction

Microscopic polyangiitis (MPA) is a small-vessel necrotizing vasculitis associated with anti-neutrophil cytoplasmic

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antibodies (ANCA) and presents itself with glomerulonephritis and hemorrhagic pulmonary capillaritis. MPA can involve any organ, but the most commonly involved organs are kidneys and lungs. The involvement of joints, skin, upper airways, and peripheral nervous system is also common [1]. Renal involvement can be seen in 80-100 % of patients and present itself by rapidly progressive glomerulonephritis [1]. The hallmark pathologic finding of MPA is pauci-immune, focal, segmental and necrotizing glomerulonephritis, which causes crescent formation in 90 % of patients [1]. The classic pulmonary involvement of MPA is diffuse alveolar hemorrhage which is reported in 12-55 % of patients [2]. ANCA is not a fixed laboratory finding and can be detected in only 50-75 % of MPA patients [1]. Peripheral nervous system involvement is common in antineutrophil cytoplasmic antibodies-associated vasculitis, but brachial plexopathy is unusual. To the best of our knowledge, no report exists about brachial plexus involvement in MPA. In what follows we present a case of MPA complicated with bilateral brachial plexopathy.

Case report

A 22-year-old man with known microscopic polyangiitis (MPA) was admitted with dyspnea and hemoptysis in March 2015. He had sought medical attention 18 months prior to his admission because of cough, dyspnea, and hemoptysis. Abdominal pain, bloody diarrhea, polyarthritis, and palpable purpura in the lower extremities were the other manifestations of his disease at the time. Table 1 shows his basic laboratory data. Blood cultures were negative. Echocardiography was normal. Chest CT scan showed ground glass pattern in the right middle lobe and lower lobes of lungs (Fig. 1). Bronchoscopy revealed bright hemorrhage



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Table 1 Laboratory values of the patient

Laboratory parameters	Normal range	Baseline (January 2014)	On admission (March 2015)
Blood			
Hemoglobin (mg/dL)	13–17	9.5	7.9
WBC ($\times 103/\mu L$)	4–10	12.3	5.4
PLT ($\times 103/\mu$ L)	100-300	346	135
ESR (mm/h)	10-20	37	39
Urine			
Protein	(-)	2+	(-)
Blood	(-)	2+	(-)
RBC (count/μL)	0–3	12–15	2
WBC (count/µL)	0–3	5–6	0
Dysmorphic RBC (%)	0	60 %	0
24-h urine protein (mg)	100-300	1500	126
Serum			
Creatinine (mg/dL)	0.8-1.2	1.2	1.1
C-reactive protein (mg/dL)	<10	35	42.8
SGOT	0-35	32	30
SGPT	0-35	28	24
Albumin (g/dL)	3.5–5	4.2	3.9
Globulin (g/dL)	2.5-3.5	3.1	3.4
Electrolytes		Normal	Normal
Immunological tests			
C-ANCA (U/mL)	<18	4.2	5.9
P-ANCA (U/mL)	<18	6.3	8.2
C3 (mg/dL)	80-120	92	85
C4 (mg/dL)	10–40	34	27
Cryoglobulin		(-)	(-)
Other auto antibodies (RF, ANA, anti-dsDNA, anti-CCP, LA, acL, anti-SSA, anti-SSB, anti-GBM)		(-)	(-)
Antibody against CMV, EBV, hepatitis B, hepatitis C, and HIV viruses		(-)	(-)

WBC white blood cells, PLT platelets, ESR erythrocyte sedimentation rate, RBC red blood cells, CRP C-reactive protein, C-ANCA cytoplasmic anti-neutrophil cytoplasmic antibody, P-ANCA perinuclear anti-neutrophil cytoplasmic antibodies, C3 complement C3, C4 complement C4, RF rheumatoid factor, ANA autoantibody to nuclear antigen, anti-dsDNA anti-double-stranded DNA, anti-CCP anti-citrullinated C peptide, LA lupus anticoagulant, acL anticardiolipin, anti-GBM anti-glomerular basement membrane, CMV cytomegalovirus, EBV Epstein–Barr virus, HIV human immunodeficiency virus, (–) negative or not detectable



Fig. 1 Diffuse bilateral alveolar hemorrhage



in the right middle lobe and lower lobes. Broncho alveolar lavage (BAL) was performed. Microbiologic examination of BAL fluid was negative for microorganisms. BAL fluid cytology showed hemosiderin-filled macrophages. Biopsy of the skin lesions showed leukocytoclastic vasculitis (Fig. 2). He was treated with methylprednisolone pulse, plasmapheresis and prednisolone 60 mg/d and cyclophosphamide 100 mg/d. His clinical symptoms were resolved and repeated CT scan after 14 days showed resolution of ground glass pattern. He was diagnosed with MPA for the following reasons: multiorgan involvement (nephritis, alveolar hemorrhage, arthritis, enteritis, and purpura), absence of autoantibodies, ruling out of infectious diseases, and leukocytoclastic vasculitis on the skin biopsy. He was

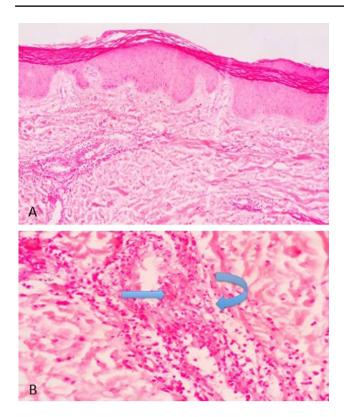


Fig. 2 Skin biopsy. a Leukocytoclastic vasculitis, **b** infiltration of vessel wall by fragmented neutrophils nuclei (*arrow* and *curved arrow*)

followed in the vasculitis clinic of Tabriz University of Medical Sciences. He had a severe flare up of the disease presented with alveolar hemorrhage 11 months ago. Up to 6 days before the admission his disease was in remission and he was on prednisolone 10 mg/d and cyclophosphamide 100 mg/d. However, he developed cough, dyspnea, and hemoptysis which increased in 6 days and had pain and paresthesia in both upper limbs. He was admitted in the ICU. On admission, he was ill and in respiratory distress. His respiratory rate was 32, and his other vital signs were in normal range. In his chest examination, coarse crackles existed in both lung fields. Neurologic examination revealed hypoesthesia, absence of deep tendon reflexes, and decreased muscle strength in the upper limbs. Because of hypoxemia and respiratory distress, he was intubated and mechanical ventilation was started. Table 1 shows his laboratory data. His chest CT scan showed ground glass pattern in the middle and lower lobes. Bronchoscopy showed previous evidence of hemorrhage without any active bleedings in the right middle and lower lobes. Treatment with imipenem, vancomycin, methylprednisolone pulse and plasmapheresis with fresh frozen plasma was started. Methylprednisolone pulse continued for 3 days and after that prednisolone 75 mg/d was started. Cytological examination of his bronchial washing material showed multiple

hemosiderin-laden macrophages. Microbiologic examination of his bronchial washing was negative for pathogenic organisms. Electromyography and nerve conduction study (EMG-NCV) showed bilateral brachial plexopathy with involvement of all of the cervical roots that were more severe in the lower roots and left side. After 16 days and 15 sessions of plasmapheresis, his symptoms improved and control chest CT scan demonstrated resolution of ground glass pattern. Therefore, mechanical ventilation and plasmapheresis were discontinued and he was extubated. Magnetic resonance imaging (MRI) of brain and cervical spine were normal. Finally, the patient was discharged with good pulmonary function, while he had severe hypoesthesia and weakness in the proximal and distal of both upper limbs, which were more severe in the left side, and atrophy of thenar and interosseous hand muscles.

Discussion

Nervous system is involved in 37–72 % of patients with MPA [2]. Peripheral nervous system involves more than central nervous system [2]. Mononeuritis multiplex and distal asymmetrical polyneuropathy are the main peripheral nervous system involvements [3]. Peripheral neuropathy usually occurs 2–16 months after the onset of vasculitis [4].

Brachial plexopathy presents itself with pain and paresthesia, sensory loss, weakness, and loss of tendon reflexes in the regions innervated by nerves in the C5-T1 segmental distribution. Trauma is the most common cause of brachial plexopathy [5]. Other causes of brachial plexopathy are paralytic brachial neuritis (Parsonage-Turner syndrome) [6], hereditary brachial plexopathy (hereditary neuralgic amyotrophy) [7], neoplasms especially small cell lung cancer, breast cancer, and lymphoma [8], radiation-induced brachial plexopathy [8], thoracic outlet syndrome [9], diabetes [10], and iatrogenic brachial plexopathies [5]. Rarely inflammatory and autoimmune diseases cause brachial plexopathy. Plexopathy in vasculitis was also observed, but it was an unusual symptom. To the best of our knowledge, we identified all the studies on brachial plexopathy in patients with systemic vasculitis by a systematic search and hand search of the literature and electronic databases, including MEDLINE, EMBASE and Google scholar for studies published between 1990 and December 2015. The search terms were: "brachial plexopathy" OR "brachial neuritis", AND "vasculitis" OR "Microscopic polyangiitis". The search resulted in 13 case reports of brachial plexopathy in systemic vasculitis. Brachial plexopathy was reported in polyarteritis nodosa, Behcet's disease, giant cell arteritis (GCA), hypersensitivity vasculitis and Henoch-Schönlein purpura [12-23]. GCA is the most common vasculitis reported as a cause of brachial plexopathy. The



Pressure of subclavian

At the course of

Unilateral

Mucosa, skin, GI tract

disease

artery aneurysm

Pressure of subclavian

At the course of

Unilateral

Mucosa, GI tract

2 years

Male

38

Behcet's disease

Lee et al. [22]

purpura

12

ī

Male

39

Behcet's disease

Yoo et al. [23]

disease

At the course of

disease

artery aneurysm

inflammatory thickenschemia secondary to Arteritis of the epineuing of the intima of the subclavian and Dissection of aorta brachial vessels Undetermined Undetermined Undetermined Undetermined Undetermined Undetermined Undetermined Undetermined Undetermined rial vessels Mechanism nitial presentation Initial presentation Initial presentation Initial presentation Initial presentation Initial presentation Initial presentation At the course of disease At the course of Time of brachial At the course of At the course of disease disease neuritis Type of brachial Unilateral Unilateral Unilateral Unilateral Unilateral Unilateral Bilateral Bilateral Bilateral Bilateral Bilateral Bilateral neuritis Lung, kidney, skin, GI Respiratory tract, skin, Skin, GI tract, joints, Skin, joints, GI tract Small and medium Other involved Large arteries size arteries tract, joints GI tract kidney organs before plexopathy Disease duration 18 months Female 6 weeks 7 days 3 days 2 days 1 Female Female Female Female Male Male Male Male Male Male Male Sex Age (years) 22 78 46 99 4 73 67 59 64 53 Polyarteritis nodosa Henoch-Schönlein Henoch-Schönlein Microscopic poly-Giant cell arteritis Giant cell arteritis Type of vasculitis Hypersensitivity vasculitis angiitis purpura Chowdhry et al. [13] London et al. [14] Dierckx et al. [15] Naseri Alavi et al. Shapiro et al. [18] Saidha et al. [17] Yilmaz et al. [20] Allian et al. [11] Blaise et al. [16] Rivest et al. [19] Agras et al. [21] Raz et al. [12] References

GI system gastrointestinal system



 Pable 2
 Summary of published cases of brachial neuritis in systemic vasculitis

brachial plexopathy in GCA is usually unilateral and occurs early in the course of disease. Table 2 shows the features of the described patient and the other cases of brachial plexopathy in the systemic vasculitis. The most acceptable explanation for the involvement of brachial plexus in vasculitis is ischemia of the subclavian artery or brachial vessels leading to plexus ischemia [16]. The other potential mechanism is direct pressure of arterial aneurysm on brachial plexus. To the best of our knowledge, brachial plexopathy has not been reported in MPA and other ANCA-associated vasculitis.

Conclusion

Peripheral nervous system involvement is common in ANCA-associated vasculitis, but brachial plexopathy is unusual. In this article, we reported a case of brachial plexopathy associated with MPA.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no potential conflict of interest.

References

- Chung SA, Seo P (2010) Microscopic Polyangiitis. Rheum Dis Clin North Am 36(3):545–558
- Collins CE, Quismorio FP Jr (2005) Pulmonary involvement in microscopic polyangiitis. Curr Opin Pulm Med 11(5):447–451
- Cattaneo L, Chierici E, Pavone L et al (2007) Peripheral neuropathy in Wegener's granulomatosis, Churg-Strauss syndrome and microscopic polyangiitis. J Neurol Neurosurg Psychiatry 78(10):1119–1123
- Cattaneo Luigi, Chierici Elisabetta, Pavone Laura et al (2007) Peripheral neuropathy in Wegener's granulomatosis, Churg-Strauss syndrome and microscopic polyangiitis. J Neurol Neurosurg Psychiatry 78:1119–1123
- 5. Wilbourn AJ (2007) Plexopathies. Neurol Clin 25(1):139-171
- van Alfen N, van Engelen BG (2006) The clinical spectrum of neuralgic amyotrophy in 246 cases. Brain 129(Pt 2):438–450

- Hannibal MC, Ruzzo EK, Miller LR et al (2009) SEPT9 gene sequencing analysis reveals recurrent mutations in hereditary neuralgic amyotrophy. Neurology 72(20):1755–1759
- Kori SH (1995) Diagnosis and management of brachial plexus lesions in cancer patients. Oncology (Williston Park) 9(8):756–760
- 9. Ferrante MA (2012) The thoracic outlet syndromes. Muscle Nerve 45(6):780–795
- Dyck PJ, Windebank AJ (2002) Diabetic and nondiabetic lumbosacral radiculoplexus neuropathies: new insights into pathophysiology and treatment. Muscle Nerve 25:477–491
- Allian SG, Towla AMH, Smith CC, Dwnie WA, Clark CJ (1982)
 Painful brachial plexopathy: an unusual presentation of polyarthritis nodosa. Postgrad Med J 58:311–313
- Raz I, Leitersdorf E, Kleinman Y (1985) Acute bilateral brachial plexus neuritis associated with hypersensitivity vasculitis. A case report and review of literature. Klin Wochenschr 63(14):643–645
- Chowdhry IA, Sinha J, Barland P (2002) Brachial plexopathy as a presenting symptom of giant cell arteritis. J Rheumatol 29(12):2653–2657
- London F, Pothen L, Lambert M, Ivanoiu A (2015) Bilateral brachial plexopathy as the presenting symptom of giant cell arteritis. Acta Neurol Belg 115(2):203–205
- Dierckx RA, Ebinger G, Herregodts P et al (1990) Recurrent brachial plexus neuropathy and giant cell arteritis. Clin Neurol Neurosurg 92(1):71–74
- Blaise S, Liozon E, Nadalon S, Vidal E (2005) Horton's disease revealed by brachial C5 plexopathy. Rev Med Interne 26(7):578–582
- Saidha S, Mok TH, Butler M, Keohane C, Harrington H (2009) Bilateral brachial neuropathy and acute aortic dissection due to giant cell arteritis. J Neurol 256(8):1346–1348
- Shapiro L, Medsger TA Jr, Nicholas JJ (1983) Brachial plexitis mimicking C5 radiculopathy-a presentation of giant cell arteritis. J Rheumatol 10(4):670–671
- Rivest D, Brunet D, Desbiens R, Bouchard JP (1995) C-5 radiculopathy as a manifestation of giant cell arteritis. Neurology 45(6):1222–1224
- Yilmaz C, Caksen H, Arslan S et al (2006) Bilateral brachial plexopathy complicating Henoch-Schönlein purpura. Brain Dev 28(5):326–328
- Agras PI, Guveloglu M, Aydin Y, Yakut A, Kabakus N (2010) Lower brachial plexopathy in a child with Henoch-Schönlein purpura. Pediatr Neurol 42(5):355–358
- Lee KY, Sunwoo IN, Oh WS, Kim SM, Choi BO (1999) Brachial plexopathy caused by subclavian artery aneurysm in Behcet's disease. Muscle Nerve 22(12):1721–1723
- Yoo WH, Kim HK, Park JH, Park TS, Baek HS (2000) Mediastinal mass and brachial plexopathy caused by subclavian arterial aneurysm in Behcet's disease. Rheumatol Int 19(6):227–230

