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Microscopic Polyangiitis Presenting with Uncommon Reversed Halo Sign: A Case Report

Nadir Görülen Ters Halo İşareti ile Ortaya Çıkan Mikroskopik Polianjitis: Olgu Sunumu

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Abstract

This case report presents an uncommon manifestation of microscopic polyangiitis (MPA) characterized by an initial reversed halo sign (RHS) on chest computed tomography (CT). A 49-year-old female patient with a history of sensorineural hearing loss presented with progressive respiratory symptoms. Comprehensive clinical investigations, including chest CT, laboratory tests and renal biopsy, confirmed a diagnosis of MPA, and targeted treatment with glucocorticoids and cyclophosphamide resulted in significant improvement in the lung lesions. This report aims to increase clinical awareness of atypical presentations of MPA and to reduce potential diagnostic challenges.

Keywords: *Microscopic polyangiitis, Reverse halo sign, tomography.*

Öz

Bu olgu sunumu, akciğer bilgisayarlı tomografisinde ilk olarak ters halo belirisi ile özdeşleşen, mikroskopik poliyanjitis (MPA)'nin nadir bir manifestasyonunu sunmaktadır. Sensorineural işitme kaybı geçmişi olan 49 yaşındaki bir kadın hasta, ilerleyici solunum sistemi semptomları nedeniyle kabul edildi. Akciğer tomografisi, laboratuvar testleri ve böbrek biopsisi dahil kapsamlı klinik tetkikler, MPA tanısını doğruladı. Glukokortikoidler ve siklofosfamid ile hedeflenmiş tedavi, akciğer lezyonlarındaki önemli bir iyileşmeye neden oldu. Bu yazı, klinisyenlerin MPA'nın atipik sunumuna olan duyarlılığını arttırmak ve potansiyel tanı zorluklarını azaltmak amacıyla hazırlanmıştır.

Anahtar Kelimeler: *Mikroskopik poliyanjitis, Ters halo belirisi, tomografi.*

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Microscopic polyangiitis (MPA) is an autoimmune disease that primarily affects small blood vessels and can cause damage to multiple organs, including the lungs and kidneys. The manifestation of the reversed halo sign on chest CT is commonly seen in various conditions, including both infectious and non-infectious diseases (1,2). To the best of our knowledge, the reversed halo sign is an uncommon manifestation in microscopic polyangiitis (MPA). This article reports a case of MPA with reversed halo sign as an initial manifestation to increase clinical awareness of the condition.

CASE

A 49-year-old housewife with sensorineural hearing loss was admitted to hospital with intermittent cough for 1 month, aggravated by expectoration for days. She had developed a mild cough 1 month earlier with occasional small amounts of white sputum after a cold that did not improve significantly with intermittent cephalosporins and levofloxacin. Her symptoms worsened over the 2 days prior to presentation with yellow sputum that was difficult to expectorate, and occasional chest tightness and shortness of breath after coughing and activity. On the day of admission she had a fever with a self-measured temperature of 38°C.

Blood routine: WBC; 7.2×10^9/L, N 81.8%, RBC; 3.7×10^{12}/L, HGB; 10.6g/L, PLT; 391×10^9/L.

Urine routine: WBC 70 cells/μL, RBC 353.7 cells/μL, urine protein (2+).

Blood gas analysis: pH 7.46, PaO_2 76 mmHg, $PaCO_2$ 32.5 mmHg, BE 0.2 mmol/L, SaO_2 96%.

Other: Liver and kidney function, myocardial enzymes, electrolytes, coagulation and blood lipids were normal. C-reactive protein was 67.60 mg/L, antistreptolysin O test was 35.0 IU/ml, rheumatoid factor was 36.3 IU/ml, erythrocyte sedimentation rate was 114 mm/h, D-dimer was 2.42 μg/ml, Widal and Weil-Felix tests were negative, and the nine components of the respiratory tract were normal. Sputum culture was negative for fungi.

Chest CT revealed multiple reversed halo signs in both lungs (Figure 1).

Painless bronchoscopy: The airway lumen remained patent, while a small amount of white and grey purulent discharge was observed (Figure 2). Transbronchial lung biopsies (TBLB), brushings and bronchoalveolar lavage (BAL) were performed in the posterior and anterior segments of the superior lobe of the right lung targeting the areas with relatively prominent lesions.

Lung bronchoscopic biopsy specimen pathology: Small tissue samples taken from the upper right and lower lung

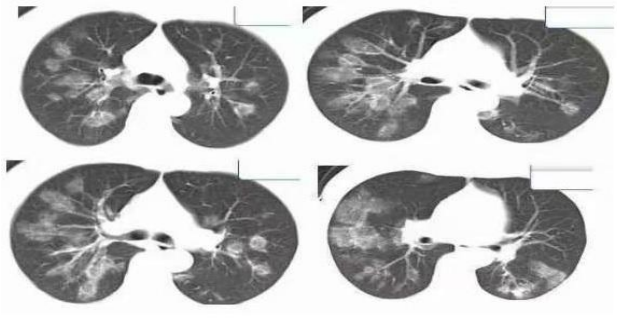

Figure 1: *Chest CT showing multiple RHS in both lungs*

Figure 2: *Bronchoscopic appearance*

bronchial mucosa were stained with hematoxylin and eosin (H&E) and sent for laboratory examination, revealing interstitial fibrous tissue hyperplasia, mild chronic inflammatory cell infiltration and a small amount of fibrinous exudation on the surface. CD56 (-), Ki67 (+ 2%), Napsin A (-), TTF-1 (-) Special staining: PAS negative (-) (Figure 3).

A microbiological examination of bronchoalveolar lavage fluid (BALF) revealed no bacteria, no acid-fast bacilli and no fungi and brush. BALF: Mycobacterium tuberculosis nucleic acid test < 500 copies/ml, GM test (-).

The patient was started initially on anti-infective treatment with moxifloxacin, with unsatisfactory results. Further pathological examinations of the lung biopsy specimen revealed mild hyperplastic interstitial fibrous tissue, a small amount of chronic inflammatory cell infiltration and a small amount of fibrinous exudation on the surface. A BALF pathogen test showed no evidence of bacteria, acid-fast bacilli or fungi, while serum anti-myeloperoxidase antibody (MPO-Ab), perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA) and anti-cardiolipin antibodies (ACA) tests were positive. A multidisciplinary panel could not exclude ANCA-associated vasculitis, and so a renal biopsy was performed, the resulting pathology of which was consistent with ANCA-associated vasculitis renal injury pointing to a final diagnosis of MPA (Table 1).

After the diagnosis was confirmed, the patient was started on oral prednisone 30mg qd, and chest CT 1 week later revealed a significant resorption of the lesions. Cyclophosphamide was added to the treatment protocol 2 weeks later (Figure 4). The patient was subsequently lost to follow-up.

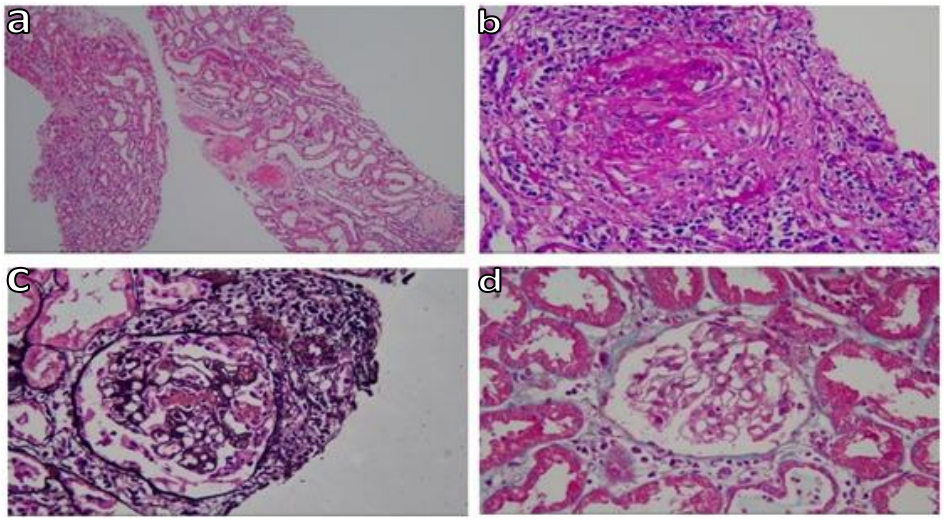

Figure 3: Dilation of some renal tubules with thickened basement membranes observed following H&E staining (a); cellular fibrous crescents identified under PAS staining (b); Small cellular crescents evident under PASM staining (c); no significant deposition of birefringent material within the glomeruli identifiable following Masson staining (d)

Table 1: Application of 2022 ACR/EULAR Classification Criteria for Microscopic Polyangiitis

Diseases	Details	Score
Criteria	**Nasal involvement:No symptoms reported**	**N/A**
Laboratory,Imaging, and Biopsy Criteria		
1. Positive test for perinuclear ANCA (pANCA) or anti-MPO antibodies	MPO-ANCA positive (1:320)	+6
2. Fibrosis or interstitial lung disease on chest imaging	Imaging shows UIP pattern indicating fibrosis	+3
3. Pauci-immune glomerulonephritis on biopsy	Proteinuria and elevated renal function consistent with pauci-immune GN	+3
4. Positive test for cytoplasmic ANCA (cANCA)	No results reported for cANCA	-1
5. Blood eosinophil count ≥ 1 x 109/L	No eosinophil count reported	-4
Total Score	**Sum of scores from above criteria**	**7**

Total Score: 7 points Diagnostic Threshold: ≥5 points

DISCUSSION

Microscopic polyangiitis (MPA) is a form of small-vessel necrotizing vasculitis that frequently affects multiple organ systems, particularly the lungs and kidneys (1,2). RHS is generally characterized by a central ground-glass opacity surrounded by a complete or partial ring of consolidation (3–5). The typical pulmonary manifestations of MPA include conventional HRCT patterns such as reticular opacities (75%) and traction bronchiectasis (38.3%) (2). To the best of our knowledge, its occurrence in MPA, as seen in our patient, is uncommon (6–8), being classically linked to cryptogenic organizing pneumonia (COP) and fungal infections (3–5,7).

In the presented case, histopathological correlation revealed a UIP-like pattern with perivascular inflammatory infiltrates and fibrosis, distinguishing it from the characteristic fibroblastic plugs seen in COP (4). This divergence underscores the critical role of histopathology in elucidating RHS's underlying etiology, especially when imaging findings overlap with COP (3–6,8).

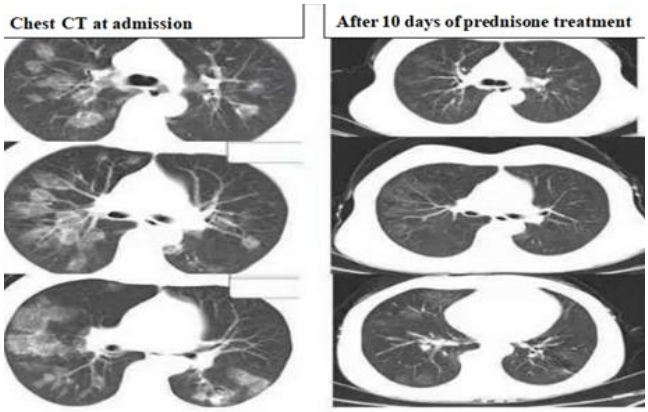

Figure 4: Chest CT at admission (first colon), and after 10 days of prednisolone treatment (second colon)

The pathogenic mechanisms underlying RHS in MPA differ fundamentally from COP's localized organizing pneumonia. ANCA-mediated vascular endothelial injuries likely trigger alveolar hemorrhage and the subsequent repair responses (9), resulting in the unique radiological-pathological discordance observed. This hypothesis aligns

with the patient's systemic manifestations and rapid treatment response to cyclophosphamide, which are atypical in classical COP management.

Renal biopsy confirmed the MPA diagnosis in the presented case (1,2), while hematuria and proteinuria further strengthened the diagnosis, along with the presence of anti-myeloperoxidase antibodies (MPO-ANCA) and perinuclear ANCA (p-ANCA), histopathological findings of interstitial fibrosis and inflammatory infiltrates (1,2), and the 2022 American College of Rheumatology/European Alliance of Associations rheumatology classification criteria for microscopic polyangiitis (10). The significant improvement noted in the pulmonary lesions following immunosuppressive therapy with glucocorticoids and cyclophosphamide corroborated the diagnosis and underscored the critical role of early therapeutic intervention (11). The lesions rapidly resolved following immunotherapy, which is atypical for infectious pulmonary pathology.

In conclusion, the identification of RHS in MPA is unusual, and so underscores the importance of expanding the differential diagnosis of RHS to include MPA, especially in cases of multi-organ involvement.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - S.W., H.W., R.J., T.L., G.L.; Planning and Design - S.W., H.W., R.J., T.L., G.L.; Supervision - S.W., H.W., R.J., T.L., G.L.; Funding -; Materials -; Data Collection and/or Processing - T.L., G.L., R.J.; Analysis and/or Interpretation - R.J., T.L., G.L.,S.W.; Literature Review - S.W., H.W.; Writing - S.W.; Critical Review - S.W., H.W.

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