ISSN: 2837-4835

Case Report

International Internal Medicine Journal

# Ulnar Nerve Palsy as the Initial Presentation of Small Vessel Vasculitis With a Negative Antineutrophil Cytoplasmic Antibody Level

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**Submitted:** 2023, Dec 29; **Accepted:** 2024, Jan 19; **Published:** 2024, Jan 30

**Citation:** Aponso, T., Wanninayaka, W. M. D. A. S., Ratnayake, R. M. S. K. (2024). Ulnar Nerve Palsy as the Initial Presentation of Small Vessel Vasculitis With a Negative Antineutrophil Cytoplasmic Antibody Level. *Int Internal Med J, 2*(1), 01-02

**Abstract**

*Small-vessel vasculitis is a rare disease entity that affects the arteries, arterioles, capillaries, and venules of the circulatory system. Small vessel vasculitis can cause severe small vessel inflammation and multisystem involvement. The respiratory tract and kidneys are the most commonly affected organs, and occasionally neurological manifestations can occur. But neurological manifestation as the initial presentation without other system involvement is very rare.*

*We report a case of a 16-year-old previously well female patient presenting with right lower limb distal muscle weakness associated with malaise, lethargy, and multiple joint pains for 2 weeks. On examination, she had palpable purpura below the knees with right sided ulnar nerve palsy. Her antineutrophil cytoplasmic antibody and antinuclear antibody testing was negative. A skin biopsy showed vasculitis involving small vessels. She was started on Prednisolone 60mg daily, followed by Mycophenolate Mofetil 500mg twice daily, and physiotherapy. She had a good clinical response to treatment over time.*

## 1. Introduction

Small vessel vasculitis is an uncommon disease entity, and ANCA-associated vasculitides have an incidence of 20 per million people. Small-vessel vasculitis affects the arteries, arterioles, capillaries, and venules of the circulatory system. Small vessel vasculitis can cause severe small vessel inflammation and multisystem involvement. Small vessel vasculitis can occur at any age but are commonly seen among the elderly and occur equally in both sexes. Around 40–50% of ANCA-associated vasculitides have negative ANCA levels [1]. The respiratory tract and kidneys are the most commonly affected organs, and occasionally neurological manifestations can occur.

Neurological manifestations can take the form of mononeuritis multiplex, sensory neuropathy, cranial nerve palsies and central nervous system mass lesions. Peripheral nervous system involvement is seen in up to 15% of patients and usually presents years after the diagnosis of the disease. Mononeuritis multiplex is the most common form of peripheral nerve involvement. Inflammation of the vasa nervorum leads to ischaemia, and axonal degeneration is the mechanism behind peripheral nerve involvement. Nervous system involvement is a poor prognostic marker in patients with small-vessel vasculitis [2,3].

Peripheral nerve involvement as the presenting feature without other system involvement is rare in the medical literature. We report a case of Ulnar nerve palsy as the initial presentation of ANCA-negative small vessel vasculitis without other systemic manifestations.

## 2. Case Report

A 16-year-old previously well female patient presented with an insidious onset of gradually progressive right hand weakness with numbness involving the palmar aspect of the hand for 2 weeks. She had arthralgia, malaise, and lethargy. She had an erythematous, non-itchy, non-painful rash below the knee bilaterally. She did not have other symptoms suggestive of connective tissue disease, and her respiratory, cardiovascular, gastrointestinal, or genitourinary involvement was not present. There was no recent history of upper respiratory tract infections. Her past medical, surgical, drug, and family histories were unremarkable. She has not had any blood transfusions, and there was no history in relation to intravenous drug abuse or sexual promiscuous behaviors.

On examination, she had a palpable purpuric rash below the knee bilaterally. The rest of the general examination was normal. On neurological examination, she had extension deformities at the 4th and 5th metacarpophalangeal joints and flexion deformities at the 4th and 5th interphalangeal joints. There was weakness in finger abduction and adduction, and Froment's sign was positive. Sensory loss was present over the 5th finger and the medial half of the 4th finger, which was suggestive of right-sided Ulnar nerve palsy, with the likely site of the lesion being distal to the elbow. Cranial nerve and cerebellar examinations were normal. The blood pressure was 110/70 mmHg with a regular pulse rate of 80 bpm. The rest of the cardiovascular, respiratory, and abdominal system examinations were normal. Her higher functions and other nervous system examinations were normal.

On investigation, the leukocyte count was 10 x 103/uL. The neutrophil count was 60%, and the eosinophil count was 2%. Hemoglobin was 11.5 g/dl, and the platelet count was 300 x 103 u/L. Urinalysis showed no red blood cells or albumin. Her serum

creatinine was 95 umol/L with a urea level of 5 mmol/L. Liver biochemistry and serum electrolyte levels were normal. The erythrocyte sedimentation rate was 45 mm per hour. Anti-nuclear Nuclear antibody, which was done by indirect immunofluorescent assay and was negative. The rheumatoid factor was negative. Cytoplasmic and Perinuclear Antineutrophil Cytoplasmic Antibodies (ANCA) were not detected in her serum. The chest radiograph, electrocardiogram, transthorasic echocardiogram, and abdominal ultrasound scan were normal.

She underwent a punch biopsy, and the biopsy revealed small vessel vasculitis involving capillaries in the deep dermis with prominent neutrophil infiltration. Her Hepatitis B/C and retroviral screenings were negative.

She was started on Prednisolone 1mg/kg daily, followed by Mycophenolate Mofetil 500mg twice daily, and physiotherapy. She had a good clinical response with time, and she is currently on rheumatology clinic follow-up.

## 3. Discussion

Small vessel vasculitides are categorized into anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitides and immune complex-associated vasculitides following the 2012 Chapel Hill Consensus Conference. Eosinophilic granulomatosis with polyangitis (EGPA), Granulomatosis with polyangitis (GPA), and microscopic polyangitis (MPA) are associated with anti-neutrophil cytoplasmic antibodies and are pauci-immune. Antineutrophil cytoplasmic antibody-associated vasculitis is characterized by loss of tolerance to leukocyte proteinase 3 (PR3) or myeloperoxidase (MPO) and the development of autoantibodies [1,4,5]. Granulomatosis with Polyangitis is associated with PR3 ANCA, and ANCA is negative in 40% of cases. Granulomatosis with polyangitis and eosinophilic granulomatosis with polyangitis are associated with MPO-ANCA, and ANCA is negative in 50% of cases [1].

Our patient had a vasculitic skin rash with mononeuropathy, arthralgia, and constitutional symptoms. In our initial differential diagnosis,we thought about ANCA-associated small vessel vasculitides, immune complex-mediated small vessel vasculitides, and medium vessel vasculitis like polyarthritis nodosa. Immune complex-mediated vasculitides, including Henoch Schonlein purpura, cryoglobulinemia, drug-induced disorders, and connective tissue disorders, show a leucocytoclastic vasculitis on skin biopsy where fragmented neutrophilic nuclei are present, which was not present in our patient [1].

ANCA is commonly associated with rapidly progressive glomerulonephritis and pulmonary involvement. 85% of GPA and MPA are associated with pulmonary involvement, and 77% show renal disease. Among EGPA patients, 25% are associated with renal involvement. Other organ systems can also be affected [3]. Though our patient only had mononeuropathy without other organ involvement,we have to follow her up closely because, at some stage in the disease course, renal and pulmonary involvement can occur in these patients, which is a bad prognostic factor.

## 4. Conclusion

It is important to screen for vasculitis-related conditions when a patient presents with multi-systemic involvement. At least one organ system involvement should prompt the screening for vasculitis since the sequelae of conditions would follow at some point in the disease course. Early diagnosis and early initiation of treatment would prevent the disastrous complications associated with Vasculitis.


## Acknowledgements

Not applicable

## Funding

None


## Contribution

Dr Tilan Aponso and Dr.W.M.D.A.S. Wanninayaka did the literature review and writing of the initial manuscript was done by Dr Tilan Aponso. Dr R.M.S.K. Ratnayake finalized the manuscript and gave expert opinion. All the authors read and approved the final manuscript.

## Ethical Declaration

Not applicable

## Consent for Publication

Informed written consent for publication of details was taken from the patient. Consent form can be made available to the editor on request.

## Competing Interests

Authors declare that they have no competing interests.

## Availability of Data and Materials

Not applicable

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