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Peripheral Neuropathy as Initial Presentation in Eosinophilic Granulomatosis with Polyangitis (EGPA)

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Abstract

Eosinophilic granulomatosis with polyangiitis, EGPA is a rare systemic vasculitis characterized by eosinophilic infiltration of various organs. EGPA can damage the peripheral nervous system in addition to the respiratory system. The purpose of this report is to underscore the relevance of evaluating EGPA as a differential diagnosis in patients with peripheral neuropathy, as well as the value of early detection and treatment. This case report describes a patient who developed peripheral neuropathy as the first symptom of EGPA. The patient's clinical course, diagnostic diagnosis, and therapy are discussed, highlighting the difficulties and need of early intervention in EGPA instances. This case report serves as a reminder to healthcare practitioners to evaluate EGPA in patients presenting with peripheral neuropathy symptoms, allowing for quick diagnosis and proper management.

Keywords: Peripheral Neuropathy, EGPA, Eosinophilic Granulomatosis with Polyangiitis, Autoimmune Disorder, Vasculitic Neuropathy, Immune Dysregulation, Eosinophil Activation

Introduction

Although it is uncommon, peripheral neuropathy might be the first sign of EGPA. EGPA, also known as Eosinophilic Granulomatosis with Polyangiitis, is an autoimmune condition that damages the blood vessels and tissues, causing harm to various organs and systems throughout the body [1]. Peripheral neuropathy can appear in EGPA patients in a variety of ways, including as polyneuropathy, sensory neuropathy, motor neuropathy, mononeuritis multiplex, and others. Although the precise mechanism by which EGPA causes peripheral neuropathy is still unclear, it is thought to be connected to the inflammatory and immune-mediated processes that are typical of the illness. Peripheral neuropathy may even develops before other symptoms including asthma, sinusitis, and nasal polyps [2]. Peripheral neuropathy in EGPA is diagnosed using a combination of imaging techniques, laboratory tests, and clinical observations. To assess the degree of nerve injury, inflammation, and imaging methods [3]. Immunosuppressive drugs like corticosteroids and methotrexate are frequently used in conjunction with supportive therapy to treat peripheral neuropathy in EGPA patients. To improve quality of life and reduce symptoms, this may also involve physical therapy and pain management. To avoid long-term damage and enhance overall results, it is essential to treat peripheral neuropathy in EGPA as soon as possible and effectively [4]. Healthcare professionals need to be aware of the possibility that EGPA could initially manifest as peripheral neuropathy in order to guarantee early detection and proper management. To inform treatment choices, prevent permanent harm, and enhance patient outcomes, timely diagnosis is essential [8].

Case Report

We present a 73-year-old male ex-smoker with a history of asthma and previous usage of a metered dosage inhaler (MDI) who came with right limb weakness, numbness, and a maculopapular rash over both legs. A physical examination indicated symptoms of mononeuritis multiplex and neuropathy. Nerve conduction studies revealed significantly reduced compound

muscle action potentials in the right common peroneal nerve with absent F-wave responses, no response in the left common peroneal nerve, no responses in bilateral tibial nerves, and mildly reduced compound muscle action potentials in bilateral femoral nerves. Sensory nerve conduction investigations revealed decreased sensory nerve action potentials in the right median and ulnar nerves, decreased sensory nerve action potentials in the bilateral sural nerves, and missing responses in the bilateral superficial peroneal nerves. Electromyography revealed fibrillation and positive sharp waves in the right tibialis anterior muscle, no spontaneous activity in the right vastus lateralis muscle, no spontaneous activity in the right first dorsal interosseous muscle, and a mild-moderate reduction in the recruitment pattern in the right first dorsal interosseous muscle. high serum p-ANCA levels (>100), normal serum c-ANCA levels (0.3), a Final FFS >2 indicating proteinuria and CNS involvement, 2+ proteinuria on urine analysis, and high ESR (109 mm/h), CRP (181 mg/L), and CPK (1612 U/L) values were found in the laboratory. Furthermore, radiological evaluation of the chest CT scan revealed fibrocalcific scarring in the apicoposterior segment of the left upper lobe, suggestive of old healed tuberculosis, patchy ground glass opacity in both lower lobes, more pronounced in the right lower lobe, raising the possibility of aspiration pneumonitis, and subtle ground glass opacity with mild edema in the posterior segment of the right upper lobe. The Interferon-Gamma Release Assay (IGRA) was negative.

Taken together, these findings confirm the diagnosis of EGPA with concomitant vasculitis, neuropathy, and pulmonary abnormalities.

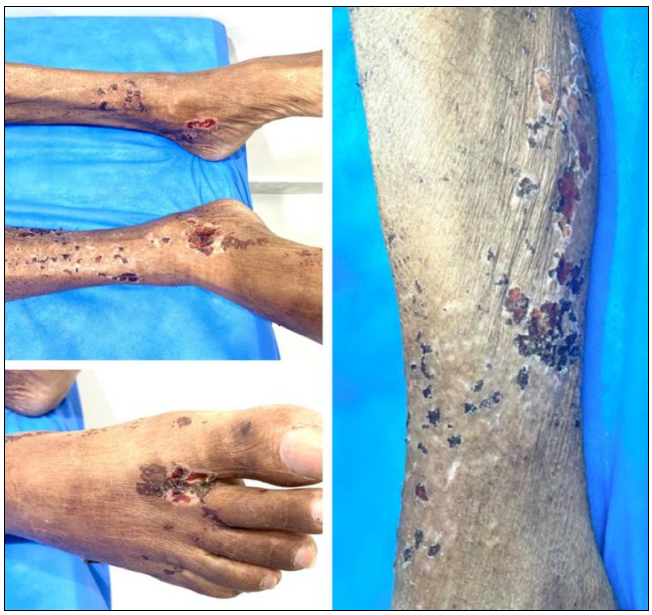

Fig 1: Vasculitic rash in a patient with eosinophilic granulomatosis with polyangiitis.

The patient was admitted to the hospital for additional treatment. To address the underlying vasculitis associated with eosinophilic granulomatosis with polyangiitis (EGPA), a regimen for cyclophosphamide induction was established. Cyclophosphamide at a dose of 1 gm was administered for a period of 6 months supplemented with Mesna in injectable form. Additionally, methylprednisolone injections were started at a dose of 1 gm for 3 days and then switched to oral high-dose corticosteroids, with a gradual tapering regime once the vasculitic rash began to improve (Fig 2).

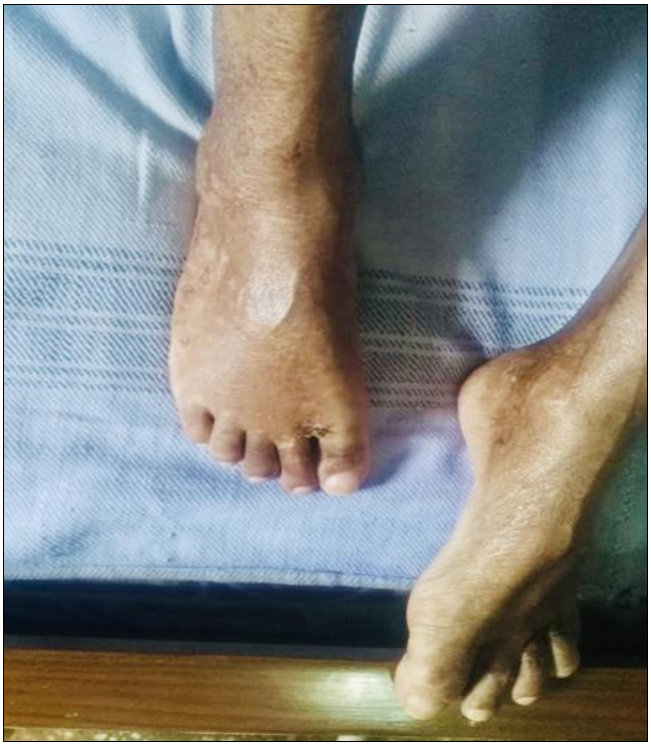

Fig 2: Notable Improvement in Vasculitic Rash

Close monitoring of the patient's clinical response and probable therapeutic adverse effects was implemented. The positive results of the treatment become evident as the vasculitis continues to improve, leading to an overall improvement in the patient's neuropathy. Remarkably, the patient regained mobility and can now walk with minimal support; this is a significant improvement over his previous wheelchair-bound state.

This case report underlines the necessity of complete evaluation, recognition of multisystem involvement, and quick intervention in the management of EGPA.

Discussion

A rare form of systemic vasculitis called eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss disease, is characterized by asthma, eosinophilia, and small-to-medium artery vasculitis. It is common for EGPA to develop as peripheral neuropathy, however this usually happens after the onset of other systemic symptoms [5]. Dr. Jacob Churg and Dr. Lotte Strauss first identified it as a condition in 1951, characterized by asthma, eosinophilia, fever, and associated vasculitis of several organ systems. In accordance with the American College of Rheumatology's diagnostic criteria from 1990, EGPA is diagnosed when at least four of the six below criteria are present [6]. Asthma, eosinophilia (>1500/dl), the onset of mononeuropathy or polyneuropathy, transient pulmonary infiltrates, paranasal sinus abnormalities, and histological evidence of extravascular eosinophils are some of the clinical criteria used to diagnosis EGPA. Elevated blood eosinophil levels are frequently detected in EGPA patients, and asthma is frequently seen in these individuals. Diagnosis can be aided by clinical, electrophysiologic, and histological investigations, including

peripheral nerve biopsies. A typical neurological EGPA manifestation is peripheral neuropathy, which can take the form of mononeuropathy or polyneuropathy. It frequently coexists with transient pulmonary infiltrates and anomalies of the paranasal sinuses. By demonstrating eosinophils outside blood vessels, histological investigation, specifically through biopsies, can aid in the confirmation of EGPA [1, 7]. As a result of mononeuritis multiplex or mixed sensory-motor peripheral neuropathy, EGPA patients often experience wrist or foot drop. The internal popliteal nerve and common peroneal nerve are most frequently affected [8]. In 75% to 80% of instances, peripheral neuropathy can be present [9]. The disease's most well-known lab characteristic is peripheral blood eosinophilia (more than 1500/dl). Additionally, 75% of patients have elevated serum IgE [10]. An ANCA positive result in EGPA is observed in about 40% of patients. Common patterns include pANCA [11]. There is currently a lack of knowledge regarding the underlying pathogenic mechanisms of vasculitic neuropathy, particularly in relation to EGPA. The interaction of humoral antibodies (ANCA), activated neutrophil granulocytes, the complement system, and endothelial cells may cause inflammation and luminal narrowing of tiny epineural arteries, which in turn causes ischemic lesions of the peripheral nerve. Immune dysregulation, eosinophil activation, and vasculitis work in concert to cause EGPA pathogenesis. White blood cells called eosinophils, which are implicated in allergy and inflammatory reactions, infiltrate different tissues and start the inflammatory cascade, which damages tissues and causes organ dysfunction [12, 15, 16]. As a result, the main cause of neuropathy is vasa nervorum blockage, which results in nerve ischemia. The loss of sensory and motor axons is the outcome of this infarction [17].

Patients with eosinophilic granulomatosis with polyangiitis (EGPA) are usually treated with a mix of drugs that are chosen based on the severity of the disease and the particular patient's characteristics. Oral prednisolone is one treatment option, with a typical starting dose of 1 mg/kg per day, followed by a slow tapering regimen. In more severe situations, an intravenous methylprednisolone pulse may be administered, followed by oral prednisolone. To maintain disease control, cyclophosphamide, an immunosuppressive medicine, may be provided at a dose of 2 mg/kg for 3 to 6 months, or cyclophosphamide pulses at a dose of 600 mg/m2 may be utilized. Methotrexate, cyclosporin A, azathioprine, and mepolizumab, a monoclonal antibody that targets eosinophils, are among other drugs that may be used. The medicine of choice will be decided by a full review by a healthcare professional based on a variety of criteria. In individuals with EGPA, these therapy options try to suppress the underlying autoimmune response, reduce inflammation, and control symptoms. Always seek the advice of a healthcare expert for individualized treatment suggestions and guidance [4, 13, 14].

Conclusion

In conclusion, whereas peripheral neuropathy as the primary presentation of EGPA is uncommon, it is critical for rapid diagnosis and appropriate care. Nasal polyps and corticosteroid responsiveness are critical diagnostic indicators. This paper investigated the clinical features, underlying mechanisms, and diagnostic techniques for this uncommon presentation. While the exact mechanisms are unknown, immune-mediated and inflammatory processes are thought to be implicated. More research is required to enhance diagnoses, therapy, and management techniques for EGPA-associated peripheral neuropathy. By raising knowledge and understanding, we can enable timely intervention and appropriate care, ultimately improving patient outcomes.

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