CASE REPOSITORY

# Low Median Nerve Palsy as Initial Manifestation of Churg-Strauss Syndrome

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Anterior interosseous nerve (AIN) syndrome is typically characterized by forearm pain and partial or complete dysfunction of the AIN-innervated muscles. Although the exact etiology and pathophysiology of the disorder remain unclear, AIN syndrome is increasingly thought to be an inflammatory condition of the nerve rather than a compressive neuropathy because the symptoms often resolve spontaneously following prolonged observation. However, peripheral neuropathy can be 1 of the first symptoms of systemic vasculitis that needs early systemic immunotherapy to prevent extensive nerve damage. Churg-Strauss syndrome (CSS; eosinophilic granulomatosis with polyangiitis) is 1 type of primary systemic vasculitis that frequently damages the peripheral nervous system. CSS—associated neuropathy usually involves nerves of the lower limb, and few studies have reported on the involvement of the upper limb alone. We report on a rare case of low median nerve palsy as the initial manifestation of CSS. The patient recovered well with early steroid treatment for primary systemic vasculitis. *(J Hand Surg Am. 2016;■(■):■—■. Copyright © 2016 by the American Society for Surgery of the Hand. All rights reserved.)*

**Key words** Anterior interosseous nerve syndrome, Churg-Strauss syndrome, systemic vasculitis, steroid treatment.

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ANTERIOR INTEROSSEOUS NERVE (AIN) syndrome is typically characterized by forearm pain and partial or complete dysfunction of the AIN-innervated muscles.[1,2] Although the exact etiology and pathophysiology of the disorder remain unclear, neuralgic amyotrophy, isolated neuritis, entrapment, vasculitis, and fascicular constriction have been considered to be associated with AIN syndrome.[1,2] Surgical exploration can be considered within 6 to 12 months of onset if the condition is considered to be a result of entrapment or fascicular constriction.[1,3] However, AIN syndrome is increasingly thought to involve an inflammatory condition of the nerve because it often resolves spontaneously following observation.[1]

Although peripheral nerve involvement is a common complication of autoimmune systemic vasculitis, owing to low incidence of the disease, little data are available on the clinical manifestations, therapy, and outcomes of vasculitic peripheral neuropathies.[4,5] Churg-Strauss syndrome (CSS; eosinophilic granulomatosis with polyangiitis) is 1 type of systemic vasculitis that is frequently associated with damage to the peripheral nervous system.[4] CSS—associated neuropathy usually involves nerves of the lower limb, and few studies have reported on the involvement of upper limbs alone.[6,7] In addition, the mononeuritis seen

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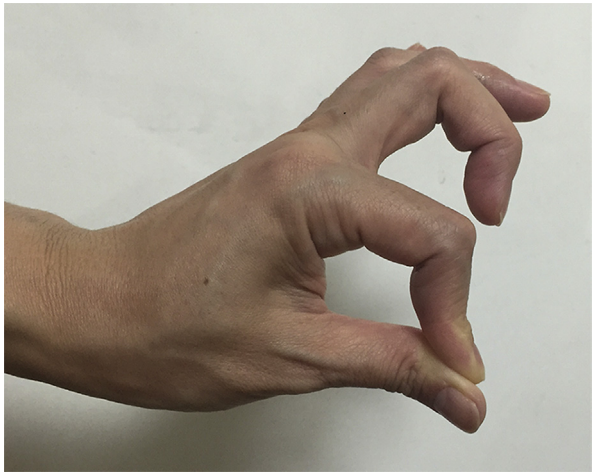

**FIGURE 1:** Photograph of the left hand shows paralysis of the flexor pollicis longus and the flexor digitorum profundus of the index finger.

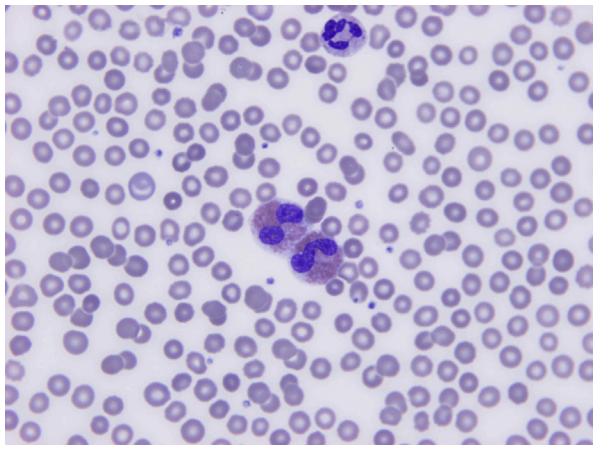

**FIGURE 2:** Photograph of the peripheral blood smear demonstrates an elevated level of eosinophil.

in CSS-associated neuropathy evolves into a polyneuropathy pattern as the disease progresses.[6,7] The recovery of vasculitic neuropathy is very slow and only partial; thus, early treatment is essential to prevent extensive nerve damage.[8]

We describe a rare case of low median nerve palsy as the initial clinical manifestation of CSS.

## CASE REPORT

A 48-year-old woman was referred by her primary care physician to evaluate a difficulty in flexing her left thumb and index finger. She also complained of a mild tingling sensation in her left thumb and index finger. She had not had any previous history of trauma to her left hand. She had a previous history of asthma, but she had no current symptoms of asthma and had not taken medication recently. There was no history of other causes of peripheral neuropathy, such as diabetes mellitus, chronic alcoholism, vitamin $B_{12}$ deficiency, or monoclonal gammopathy. A physical examination confirmed partial paralysis of the flexor pollicis longus (FPL), which was rated as grade 2 in power, and complete paralysis (grade 0 power) of the flexor digitorum profundus (FDP) of the index finger (Fig. 1). No Tinel sign or tenderness over the course of the median nerve could be detected. An electromyography performed 2 months after the onset of the weakness revealed decreased recruitment in the index FDP and pronator quadratus and no voluntary motor unit potentials from the FPL, consistent with AIN neuropathy. Laboratory tests revealed eosinophilia (13%) in the differential white blood cell count (Fig. 2). Owing to her previous history of asthma and elevated level of eosinophils in the differential count, she was referred to the department of rheumatology at our hospital for a suspected autoimmune disease.

After additional investigations, an experienced rheumatologist diagnosed her with primary systemic vasculitis based on the criteria of the American College of Rheumatology for CSS (eosinophilic granulomatosis with polyangiitis),[9] which classify CSS as the presence of at least 4 of 6 items including the history of asthma, the presence of eosinophlia (Fig. 2), development of mononeuropathy or polyneuropathy, nonfixed pulmonary infiltrates on plain radiographs (Fig. 3), paranasal sinus abnormality (Fig. 4), and biopsy showing extravascular eosinophils. Corticosteroid therapy (consisting of intravenous methylprednisolone at 1000 mg/day for 3 consecutive days before starting oral prednisone) was administered to treat her CSS. The progression of CSS was well controlled with steroid therapy alone, and no additional peripheral neuropathies were observed throughout the course. She started to recover from the palsy 4 months after the onset of palsy (about 1 month after initiating immunotherapy). Six months after the onset of palsy, her final muscle power scores were grade 5 for FPL and grade 4 for FDP of the index finger, and she had little difficulty in performing activities of daily life.

## DISCUSSION

Anterior interosseous nerve syndrome is rare and the exact pathophysiology of the disorder is unclear. Therefore, there is a lack of consensus on the timing of surgical intervention. The evidence that the AIN syndrome is generally self-limited outweighs the evidence suggesting external nerve compression that might benefit from surgery because there is a high probability of spontaneous resolution after even 1