An Unusual Cause of Swan Neck Deformity of the Fingers

P. Rajendran Srijithesh, MD, DM

Objective: To describe a patient who presented with nonfixed swan neck deformity of the fingers and generalized body aches.

Design: Case report.

Setting: Tertiary care teaching hospital.

Patient: A 38-year-old woman who presented with swan neck deformity of the fingers.

Main Outcome Measures: Electromyographic finding and electromyographic and clinical response to intravenous immunoglobulin.

Results: Needle electromyography revealed continuous motor unit activity in the 50- to 70-Hz frequency during the resting state for all the muscles sampled, which suggests the possibility of neuromyotonia. After ruling out possible secondary causes, we treated the patient with intravenous immunoglobulin, considering primary neuromyotonia. The body ache improved by 100% on the visual analogue scale, and the electromyographic discharges disappeared from the paraspinal and tibialis anterior muscles and changed in its morphology to doublets, triplets, and multiplets in the first dorsal interossei and flexor digitorum profundus.

Conclusion: Neuromyotonia should be considered in the differential diagnosis of swan neck deformity of the fingers, especially in cases that show no fixed deformity and are not associated with any other rheumatologic stigmata.


WAN NECK DEFORMITY OF THE fingers involving flexion at the distal interphalangeal joint and hyperextension at the proximal interphalangeal joint is described in the context of many rheumatologic diseases and is thought to be due to an imbalance in the flexion and extension forces acting across the finger joints.1 In early rheumatoid arthritis, the deformity can be “nonfixed” because of an imbalance of forces acting across the finger joints that is due to laxity of the ligaments.

REPORT OF A CASE

A 38-year-old woman presented with complaints of generalized body aches and flexion deformity of the fingers of both hands of 3-year duration. There was no weakness of the hand muscles, and the deformity persisted during sleep. There was no fasciculation or myokymia, nor were there any abnormal movements.

On examination, there was flexion of the distal interphalangeal joints and hyperextension of the proximal interphalangeal joints, which were most prominent on the third and fourth digits of the left hand (Figure and video, http://www.jamaneuro.com). The deformity was not fixed and could be corrected by passive movement. Apart from this, the results of her general systemic and neurological examination were normal. The results of all the relevant investigations into her condition were within the normal limits (Table).

 Needle electromyography of the flexor digitorum profundus and extensor indicis revealed continuous motor unit activity in the 50- to 70-Hz frequency during the resting state for all the muscles sampled. The motor unit action potential morphology was normal. Electromyography was performed on the first dorsal interossei, deltoid, tibialis anterior, and thoracic paraspinal muscles, and these muscles also showed continuous motor

Video available online at www.jamaneuro.com

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unit activity during the resting state. Given the absence of any other disease involving the peripheral nervous system or of any evidence of neoplastic illness, the possibility of primary neuromyotonia was considered.

Considering the possibility of an autoimmune etiology, the patient was given a course of methylprednisolone sodium succinate followed by oral prednisolone sodium phosphate. After 2 weeks, another electromyogram showed the disappearance of discharges from the tibialis anterior and paraspinal muscles, but the motor unit discharges in the extensor indicis, first dorsal interossei, and flexor digitorum profundus persisted. Pain had improved by 30% on the visual analogue scale. Subsequently, the patient was given intravenous immunoglobulin at a dose of 0.4 g/kg/d for 5 days. After 1 week, another electromyogram showed a decrease in the frequency of discharges from the first dorsal interossei, extensor indicis, and flexor digitorum profundus, with discharges occurring in doublets, triplets, and multiplets rather than as continuous motor unit discharges. No discharges were noted in the paraspinal and tibialis anterior muscles. On the visual analogue scale, pain relief was complete, and joint relaxation improved by 80%. The patient was continued on oral prednisolone, with azathioprine sodium as a steroid-sparing agent.

**COMMENT**

Neuromyotonia is a clinico-electrophysiological syndrome characterized clinically by muscle stiffness, myokymia, and pseudomyotonia and characterized electrophysiologically by neuromyotonic and myokymic discharges. The case is unusual in that it presented with focal finger flexion deformity while having generalized electrophysiological abnormality. Although patient had extensive electromyography-detected continuous motor unit activity, there were no clinically remarkable features suggestive of neuromyotonia.

Electrophysiologically, both neuromyotonic and myokymic discharges are repetitive discharges originating from motor units. They are differentiated morphologically by their frequency and cadence of occurrence. Neuromyotonic discharges occur in the high-frequency range of 150 to 250 Hz with a characteristic decrementing tempo, whereas myokymic discharges occur rhythmically as grouped fasciculations with a high intraburst frequency of 5 to 60 Hz and a slow interburst frequency (<2 Hz) characteristically producing a marching sound during electromyography. These discharges are generated by damaged peripheral motor axon, and, therefore, they do not disappear with general or spinal anesthesia, nor are they affected by supraspinal influences like sleep.

As a primary disorder, neuromyotonia results from autoantibodies directed against presynaptic voltage-gated potassium channels. Considering the presumed autoimmune etiology in primary neuromyotonia, treatment with immunomodulators like steroids and immunoglobulins have been tried in primary neuromyotonia.6,7

The focal presentation of neuromyotonia has been reported previously. Modarres et al8 reported 2 patients with restricted high-frequency continuous motor unit discharges in the flexor digitorum superficialis and extensor communis manifesting as finger flexion abnormality of the middle and ring fingers. Jamora et al9 reported a patient who presented with painless flexion restricted to the little finger that showed evidence of generalized neuromyotonic and myokymic discharges on an electromyogram. However, to my knowledge, neuromyotonia presenting with a combination of flexion and extension abnormalities in adjacent joints to produce a “swan neck” deformity of the fingers has hitherto been unreported. I hypothesize that this case may be due to a variable contraction of the extensor and flexor muscles in the finger joints, which causes the peculiar deformity.

To conclude, neuromyotonia should be considered in the differential diagnosis of swan neck deformity of the fingers, especially in cases that show no fixed deformity and are not associated with any other rheumatologic stigmata. Idiopathic neuromyotonia deserves a trial with steroids and immunoglobulins.

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**Figure:** Swan neck deformity of the fingers.

**Table. Relevant Investigations Into Swan Neck Deformity of the Fingers of a 38-Year-Old Woman**

<table>
<thead>
<tr>
<th>Investigations</th>
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<tbody>
<tr>
<td>Hemogram with erythrocyte sedimentation rate</td>
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<tr>
<td>Peripheral smear</td>
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<tr>
<td>Blood urea and serum creatinine levels</td>
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<tr>
<td>Liver function tests</td>
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<tr>
<td>Serum electrolytes analyzed (sodium, potassium, calcium including ionized fraction, magnesium)</td>
</tr>
<tr>
<td>Arterial blood gas analysis</td>
</tr>
<tr>
<td>Thyroid function test</td>
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<tr>
<td>Thyroid antibodies (antithyroid peroxidase, antithyroglobulin)</td>
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<tr>
<td>Antinuclear antibodies, anti–double-stranded DNA, rheumatoid arthritis factor, acetylcholine receptor antibody</td>
</tr>
<tr>
<td>Human immunodeficiency virus, hepatitis B surface antigen</td>
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<tr>
<td>Chest radiography, ultrasonography of the abdomen</td>
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<tr>
<td>Nerve conduction study</td>
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<tr>
<td>Routine cerebrospinal fluid study (cell count and protein and sugar levels)</td>
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*a* The results of all the relevant investigations into her condition were within the normal limits.
Correspondence: P. Rajendran Srijithesh, MD, DM, Department of Neurology, Jawaharlal Institute of Postgraduate Medical Education and Research, Dhanvanthri Nagar, Pondicherry 605006, India (srijitheshpr@rediffmail.com).

Conflict of Interest Disclosures: None reported.


REFERENCES