Frontal "wasting": Cortical arteriovenous malformation causing hand wasting

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Case Report (Clinical Description)

A 15-year-old boy with no known co-morbidities presented with gradual onset progressive thinning of the left hand associated with weakness and occasional numbness of the left hand of 4 months duration.

Motor system examination revealed disproportionate wasting in the medial compartment of left hand with partial clawing [Figure 1a]. Hand grip was weak with normal power at the shoulder and the elbow joints. Deep tendon reflexes were brisk (3+) in the left upper and lower limbs with normally elicitable reflexes (2+) on the right side. Bilateral plantar response was flexor. Rest of the systemic and neurological examination including sensory examination and cortical sensations were normal. The routine investigations of the patient were unremarkable. Sensory/motor nerve conduction study and electromyography of the wasted muscles was normal. Magnetic resonance imaging (MRI) of brain and spine were essentially normal. Neuroimaging of brain revealed multiple tortuous vascular flow voids suggestive of arteriovenous malformation (AVM) in the right posterior frontal lobe [Figure 1b and c]. Digital subtraction angiography confirmed evidence of entangled vessels suggestive of AVM at the same site [Figure 1d and e]. The patient was offered choice between embolization (preferably) and gamma knife surgery for the AVM; however, he opted for gamma knife surgery.

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Discussion

The usual neurological causes of the hand wasting are the ones caused by lower motor neuron lesions. The assumption that focal muscular atrophy is "only" caused by a lower motor neuron lesion can be misleading. Unusually, cortical lesions may cause focal amyotrophy, perplexing the clinical scenario. The evidence supporting the role of transneuronal degeneration (TND) of corticospinal fibers in such cases is convincing.

Claw hand is an abnormal hand position that develops due to wasting and weakness of the intrinsic hand muscles and sparing of the long flexor and extensor muscles of the forearm. The usual neurological causes of the hand wasting are nerve/plexus injuries, neuropathies, anterior horn cell disorders, or distal myopathies.[1]

The presence of brisk reflexes with pure motor involvement in the form of wasting raises suspicion of anterior horn cell disorder. In a young male with asymmetrically wasted hand, the possibility of "Hirayama disease"[2] or monomelic variant of motor neuron disease should also be ruled out.

Rarely cortical lesions may also produce focal wasting and weakness.[3] Lesions of parietal or the frontal cortex may present

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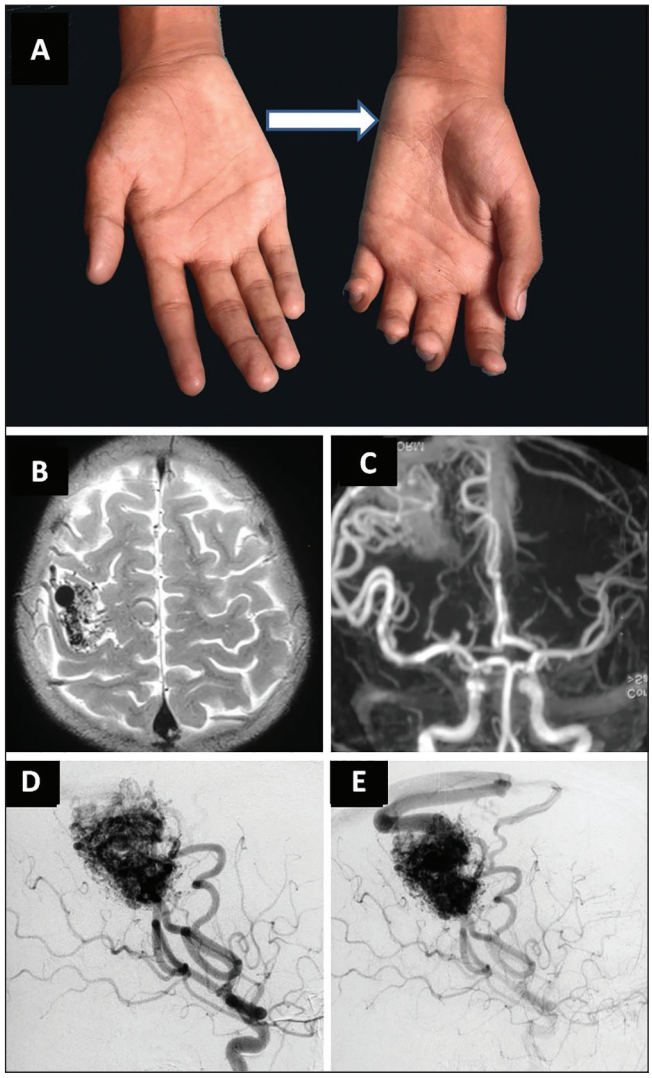

Figure 1: Showing disproportionate wasting of the hypothenar eminence of left hand with partial clawing (a) Axial T2 weighted (b) and contrast enhanced magnetic resonance angiography employing the time of flight (TOF) method (c) Reveals abnormal flow voids in the right frontal lobe suggestive of AVM. Digital subtraction angiography (d and e) Better demonstrates the AVM nidus with arterial feeders from the middle cerebral artery (MCA) branches and venous drainage into the superior sagittal sinus

with localized atrophy of hand along with brisk reflexes.[4] The proposed theories are disuse, pressure or traction neuropathies, vasomotor changes, and loss of upper motor neuron trophic influence.

In humans, the role of TND (anterograde or retrograde) is well known to occur in lesions of visual, limbic, or dentato-rubro-olivary pathways. This phenomenon is not that well established in the somatic motor system. Kondo *et al*.[5] have reported that the degree of pyramidal tract degeneration seemed to be paralleled by fiber loss in ventral spinal roots. Qui *et al*.[6] also suggested that atrophy of neurons in the cervical segment occurred on the side of lateral cortical spinal tract (CST) degeneration. In electrophysiological studies, motor units reportedly are decreased in number on the side of the spinal cord affected by cerebral stroke, with alpha motor neurons being in a functionally depressed state.[7]

Another controversial issue is the exact site of upper motor neuron lesion (UMN) lesion causing muscle wasting. Parietal "wasting" has been described as case reports and it has been insisted that parietal lobe is the sole source of trophic influence. To some, it seems more likely that it is the motor cortex that provides the trophic influence to the motor neurons.[8] Hence, TND of motor neurons may occur following lesions of the contra lateral motor cortex or emergent fibers.

Conclusion

To conclude, our case report provides further evidence that a strategically placed lesion in the cortex can be a source of isolated hand wasting. This case report also emphasizes the value of extensive and detailed clinical examination in neurological clinical practice.

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Conflicts of interest
There are no conflicts of interest.

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