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Role of Dynamic Flexion Mri In Hirayama Disease -A Case Series

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Abstract: *Hirayama disease (HD) is a benign focal amyotrophy of the distal upper limbs characterized by distal asymmetric weakness and wasting of upper extremi ties mainly affecting the lower cervical and upper thoracic segmental myotomes(C7,C8,T1) . There is sparing of the brachioradialis and proximal muscles of the upper limb innervated by C5–6 myotomes. The disease most often occurs in young adults, with a male predominance . It is mostly sporadic. . It is commonly seen in Asia and rarely encountered in the Middle East countries.The condition is belived to be caused by excessive anterior movement of cervical spinal cord on flexion,resulting in anterior compression of cord against posterior vertebral column with resultant ischemia and cord atrophy. In our study four patients with clinically suspected Hirayama disease were evaluated with neutral position, flexion, contrast-enhanced MRI and fast imaging employing steady-state acquisition (FIESTA) sequences. The spectrum of MRI features was evaluated and correlated with the clinical and electromyography findings. MRI findings of localized lower cervical cord atrophy , asymmetric cord flattening, loss of attachment of the dorsal dural sac and subjacent laminae in the neutral position, anterior displacement of the dorsal dura on flexion and a prominent epidural space were revealed in all patients on conventional MRI as well as with the dynamic 3D-FIESTA sequence. Intramedullary hyperintensity was seen in all patients on conventional MRI. Flow voids were seen in two patients on conventional MRI sequences and in all patients with the 3D-FIESTA sequence. Contrast enhancement of the epidural component was noted in all the four patients with thoracic extensions. . Hirayama disease is rarely encountered in clinical settings and should be suspected in young male patients presenting with unilateral or asymmetrical bilateral lower motor weakness of hands and forearms.*

Keywords: *Hirayama disease ,Benign focal amyotrophy, 3D-FIESTA,Flexion MRI.*

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I. Introduction:

Hirayama disease, is characterized by the insidious onset of unilateral or asymmetric oblique amyotrophy affecting the C7, C8 and T1 myotomes. It occurs in young men and is characteristically associated with cold paresis. The onset is insidious, and is characterized by muscle weakness and atrophy in the hand and forearm, with sparing of the brachioradialis, giving the characteristic appearance of oblique amyotrophy.[1] The amyotrophy is unilateral in most patients, asymmetrically bilateral in some and rarely symmetric. Hirayama disease is a benign, nonprogressive motor neuron disease. Chronic microcirculatory changes in the territory of the anterior spinal artery induced by repeated or sustained flexion account for the necrosis of the anterior horns of the lower cervical cord, which is the hallmark on pathology.[1,2] A spectrum of diagnostic magnetic resonance imaging (MRI) features has been described in the literature. The present study reviews the MRI features in Hirayama disease in the neutral and flexion positions and the use of the newer 3D fast imaging employing steady-state acquisition (FIESTA) sequence on a 1.5 Tesla MRI scanner.

II. Materials And Methods:

This was a prospective cross-sectional study conducted from January 2020 to august 2021 in a tertiary care center from south India. Four patients with clinically definite Hirayama disease underwent electrophysiologic evaluation followed by MR imaging of the cervical spine.

MR Imaging Protocol: MR imaging was performed using a 1.5T Philips acheiva MRI scanner. Image acquisition of the cervical spine was initially performed with patients in a supine neutral position in routine sagittal T2- and T1-weighted spinecho, sagittal and coronal STIR, and axial T2- and T1-weighted fast spin-echo and axial 2D T2*WI gradient recalled-echo sequences. Sagittal spin-echo T1WI was acquired with a TR/TE of 450 –500/9 –15 ms; sagittal T2WI, with a TR/TE of 4000 – 4600/ 110 –120 ms with a 3-mm slice thickness.

Axial 2D T2*WI gradient recalled-echo imaging was performed with a TR/TE of 650 – 750/24 –32 ms and a flip angle of 24°–28°. Flexion MR imaging of the cervical spine was performed with a body coil without using a cervical coil. The optimum neck flexion of 30°– 40° was obtained after putting soft MR imaging– compatible support behind the nape of neck with further support on either side of the neck to create immobility of the neck during flexion MR imaging. Post gadolinium fat-suppressed sagittal and axial T1-weighted images of the cervical spine were obtained in neck flexion with a slice thickness of 3 mm.

Clinical History:
4 male patients of 17, 21,22 and 24 years of age respectively presented with complaints of bilateral upper limb weakness, gradually increasing tremors and fasciculations in bilateral upper limbs of 2-4 months duration.Complaints were more significant in one limb than the other.On examination there is atrophy of distal muscles of both hands with sparing of brachioradialis in all the patients.

Imaging Findings:
Patients underwent MRI cervical spine with additional flexion and post-contrast imaging as per the protocol for hirayama disease.

Case 1: On Neutral MRI, there is mild to moderate cord flattening. On flexion MRI, there is flattening of the spinal cord, obvious posterior dural detachment and prominent venous flow voids on the T2-weighted image. Post-contrast images show mild homogenous post-contrast enhancement.

Case 2: On MRI, there is abnormal cervical curvature and focal atrophic changes at C6-7 level. There is a forward movement of cord with a reduced anteroposterior diameter on the flexion scan. There is a forward displacement of lower cervical vertebrae, flattening of the spinal cord and obvious posterior dural detachment.

Case 3: On neutral MRI, there is an abnormal cervical curvature and focal T2 hyperintense signal in the cervical cord. On flexion MRI, there is flattening of the spinal cord, loss of posterior dural attachment and prominent venous flow voids on T2-weighted image.

Case 4: On neutral MRI, there is an abnormal cervical curvature and focal T2 hyperintense signal in the cervical cord. On flexion MRI, there is flattening of the spinal cord, loss of posterior dural attachment and mildly prominent venous flow voids on T2-weighted image. Post-contrast images show intense homogenous post-contrast enhancement of posterior dural space.

III. Discussion:

Hirayama disease is a benign form of focal amyotrophy that affects C7, C8, and T1 segmental myotomes with sparing of the brachioradialis, giving the characteristic appearance of "oblique amyotrophy," and sparing the proximal muscles of the upper limb innervated by C5– 6 myotomes. It manifests with gradual unilateral or asymmetric bilateral muscle weakness and atrophy in young males and is usually self-limited. It can be associated with autonomic dysfunction in distal upper limbs in the form of exaggeration of weakness on exposure to cold (cold paresis), cold skin, excessive sweating, and hair loss over the dorsum of the hands, and bilateral minipolymyoclonus. Approximately 70% of the patients experience disease progression within 3 years, and approximately 95% stabilize 5 years after disease onset [3]. It differs from classical types of motor neuron diseases because of its self-limited nature and the pathological findings of chronic microcirculatory changes in the anterior horns of the lower cervical cord. The pathogenesis of Hirayama disease still remains obscure. The first pathological study was published by Hirayama et al. [4] in 1987 on a patient after 23 years of disease onset and showed necrosis and degeneration of various degrees of large and small nerve cells with mild gliosis only in the anterior horns of the spinal cord at C5 to T1, particularly marked at C7 and C8. They suggested circulatory insufficiency in the lower cervical cord as the leading cause. However, current neuroradiological techniques are able to show forward displacement of the posterior wall of the lower cervical dural canal when the neck is flexed, causing marked and often asymmetric flattening of the lower cervical cord, which is presumed to be the primary pathogenetic mechanism of Hirayama disease [5.6]. In "Hirayama disease" patients, there is imbalance in growth of the vertebrae and the dura mater leading to a "tight dural canal" and "overstretched cord," which cannot compensate for the increased length of the posterior wall during flexion. This causes an anterior shifting of the posterior dural wall, with consequent compression of the cord. This compression from repeated or sustained flexion of the neck may cause chronic microcirculatory disturbances in the anterior portion of the spinal cord may produce necrosis of the anterior horns, which are most vulnerable to ischemia [7].
Clinical diagnosis is confirmed by electrophysiological and radiological studies. EMG findings are indicative of chronic denervation noted in the C7, C8, and T1 innervated muscles, with or without acute denervation

potentials (fasciculations, positive sharp waves and fibrillations potentials). "Reverse split hand syndrome," which is characterized by decreased/absent CMAP amplitude in the abductor digiti minimi while preserved in the abductor pollicis brevis, is usually seen in nerve conduction study. It differentiates "Hirayama disease"from ALS which shows classical "Split hand syndrome" [8,9].

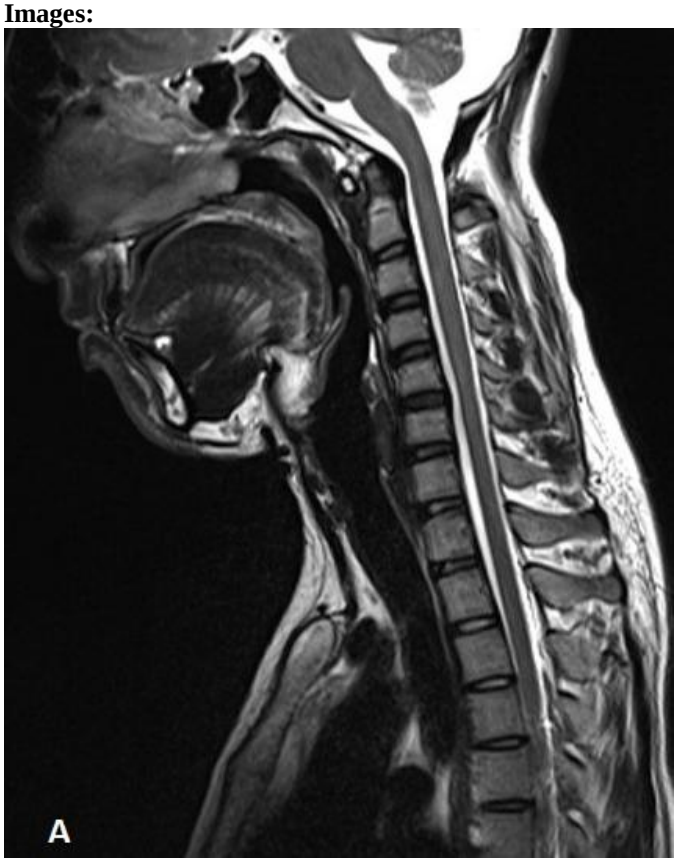
Routine MRI in neutral position is often reported as normal, but it can also show lower cervical cord atrophy or abnormal cervical curvature (straight or kyphotic). However, the classical MRI findings of cervical spine in neck flexion include: forward displacement of the posterior wall, loss of the posterior dural sac attachment with adjacent lamina, and a wellenhanced crescent-shaped mass in the posterior epidural space of the lower cervical canal. This is thought to represent congestion of the posterior internal vertebral venous plexus as it disappears on neutral neck position [6]. The increase in the laminodural space and the presence of cervical cord flattening during flexion are essential for diagnosis as described by Boruah et al. [10] in 45 patients with clinically definite "Hirayama disease." He found that the laminodural space at maximum forward shifting of the posterior dural sac ranged from 3 to 9.8 mm, with a mean distance of 5.99 mm. Vitale et al. [11] suggested an MRI protocol including: sagittal T1-weighted and T2- weighted sequences and axial T2 or T2*-weighted sequence in neutral position; sagittal T2- weighted sequences and axial T2 or T2*-weighted sequence in a neck-flexion of 25–35 degrees in addition to sagittal T1-weighted sequences in neck-flexion before and after gadolinium intravenous administration. Snake-eyes appearance, a radiological finding described as a symmetrical bilateral small high-signal-intensity lesion on axial T2-weighted MRI, appears during the late stage of "Hirayama disease" and is proposed as an indicator of irreversible damage and poor prognosis [12]. According to a nationwide study in Japan by Tashiro et al. [4], the following criteria are important for diagnosis: (1) distal predominant muscle weakness and atrophy in the forearm and hand; (2) involvement of the unilateral upper extremity in most cases; (3) onset between the ages of 10 to early 20s; (4) insidious onset with gradual progression for the first several years, followed by stabilization; (5) no lower extremity involvement; (6) no sensory disturbance or tendon reflex abnormalities; and (7) exclusion of other diseases. All these criteria were fulfilled in our patient. Several conditions can be considered in the differential diagnosis of "Hirayama disease" including: amyotrophic lateral sclerosis, spinal muscle atrophy, C8-T1 radiculopathy, compressive myelopathy, cervical spondylotic myelopathy, syringomyelia, post-polio syndrome, multifocal motor neuropathy, and toxic neuropathy [13].

As "Hirayama disease" is considered a self-limited disease and often stops progressing after 1–5 years of onset, the mainstay of treatment consists of preventing neck flexion using a cervical collar to halt further progression. Its application at an early stage of the disease for 3– 4 years has been advocated with good response [14]. EMG can be used as an indicator to start and to stop cervical collar therapy depending on changes of latency and amplitude of motor evoked potentials and persistence of F wave on neck flexion [15]. Good prognosis is seen in patients with shorter duration of illness and no or mild cord atrophy in neutral neck position [14].

IV. Conclusion:

Hirayama disease should be suspected in young males presenting with weakness,muscle atrophy,fasciculations and tremors of distal upper limbs and should undergo dynamic flexion MRI cervical spine as per protocol to exclude other diagnosis and initiate proper early treatment.This approach prevents misdiagnosis and mistreatment to the patients.

Images:



CASE 1

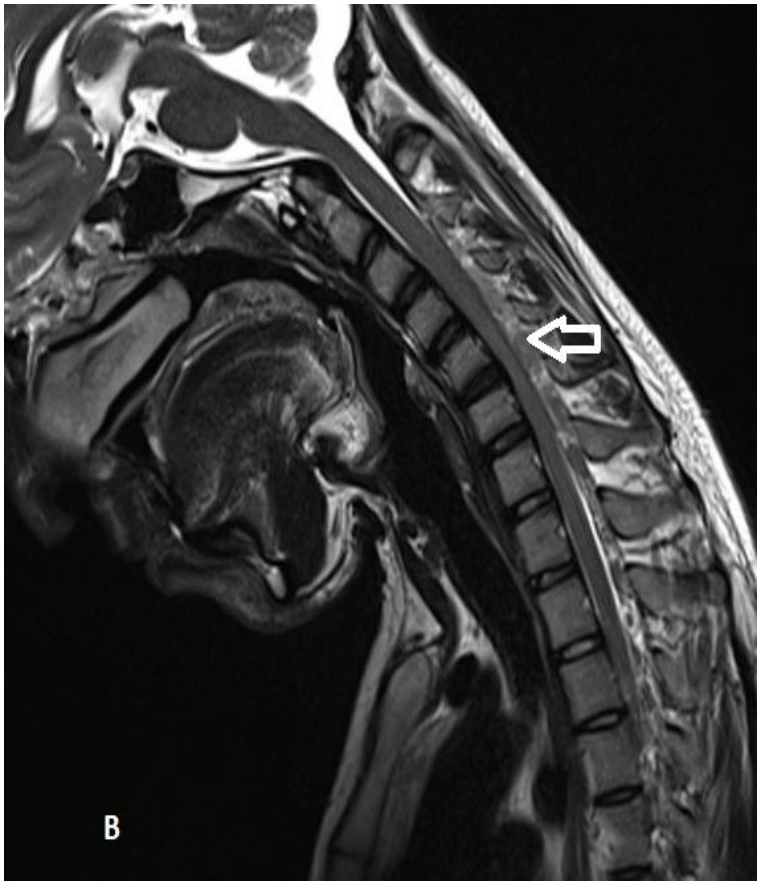


Figure 1 A,B: T2W Sagittal image in neutral position(A) demonstrates no significant abnormality. T2W Sagittal image in flexion position(B) in same patient demonstrates detachement of posterior dural sac with multiple tortuous flow voids from C4 TO T1 level.(white arrow in B)

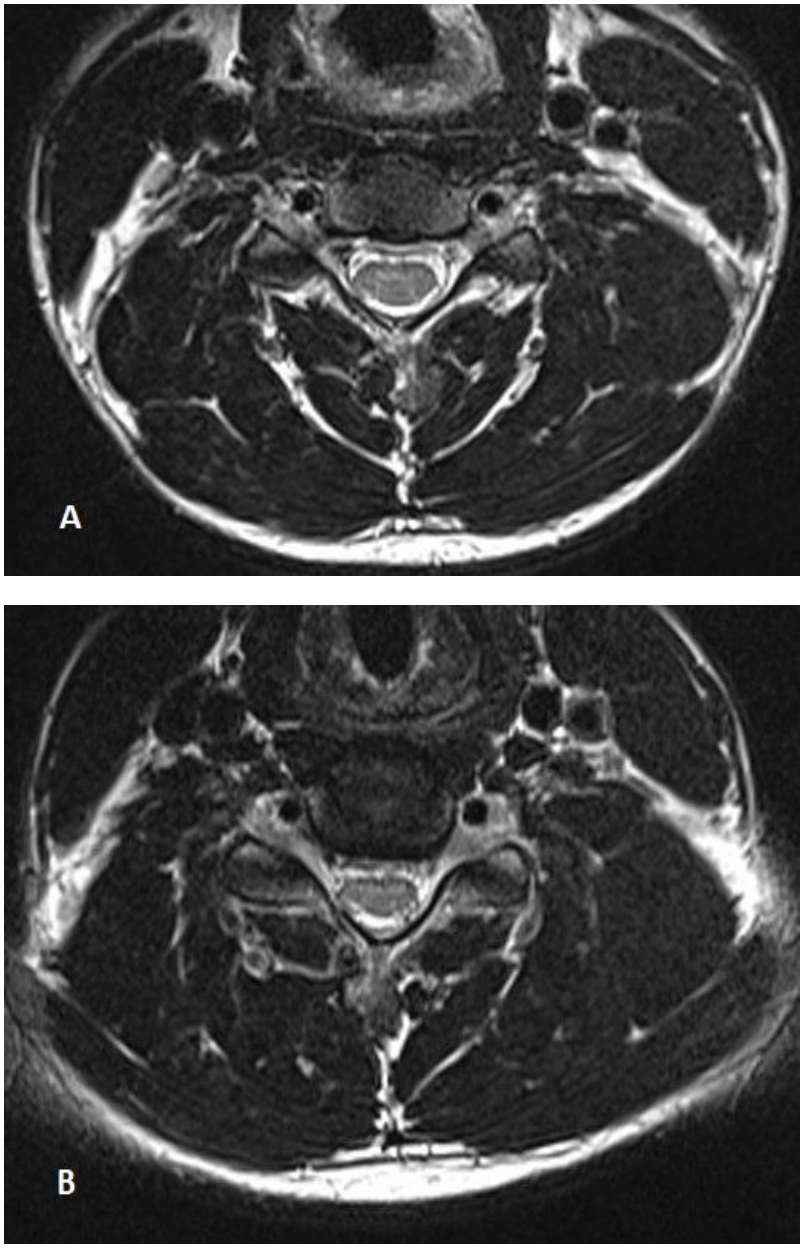


Figure 2A,B: Axial T2W image at C3 level(A) demonstrates normal cord,there is flattening of cord with associated T2 hyperintensity at C6 level(B).

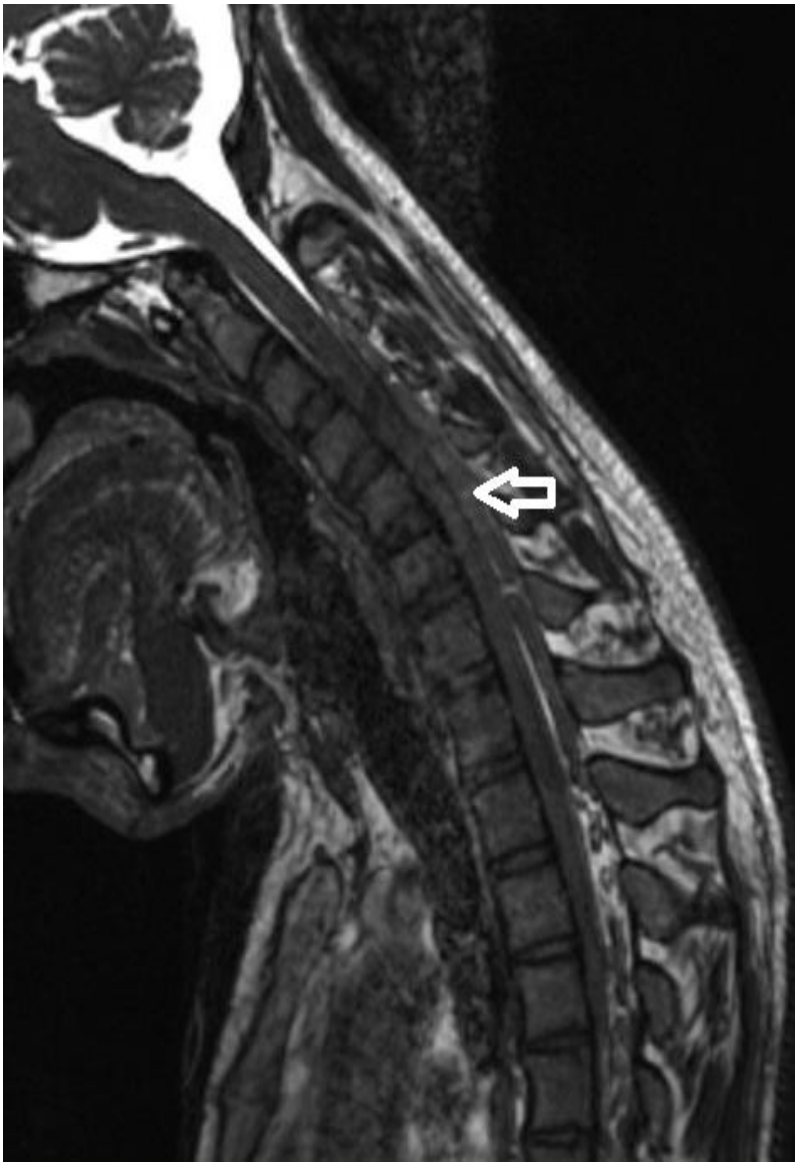


Figure 3: Sagittal **3D FIESTA** image shows detachement of posterior dural sac from C4 to T1 level with multiple flow voids within.

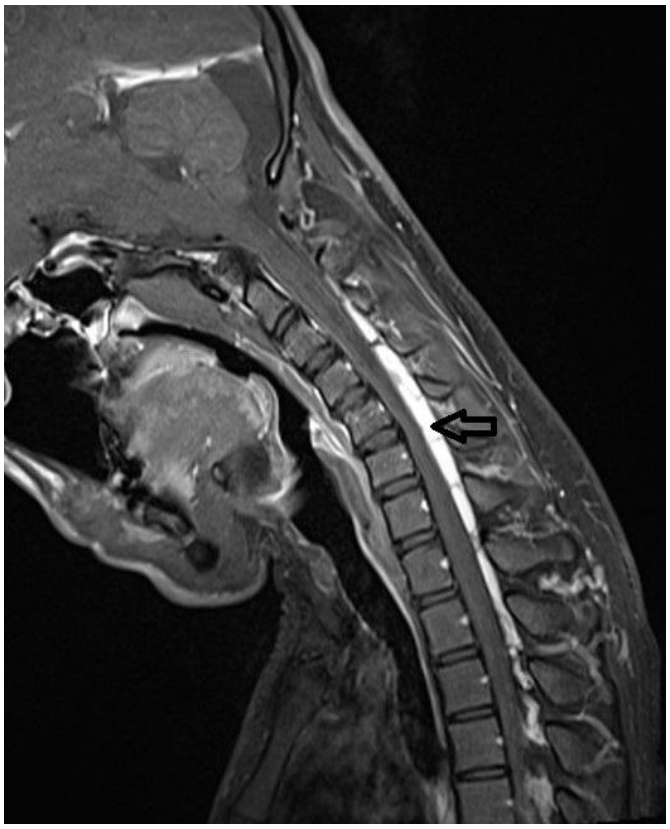


Figure 4: Sagittal T1 fat sat post contrast image in flexion position shows intense enhancement (black arrow) of the posterior dural space extending from C3 vertebtal level upto the upper thoracic vertebrae.

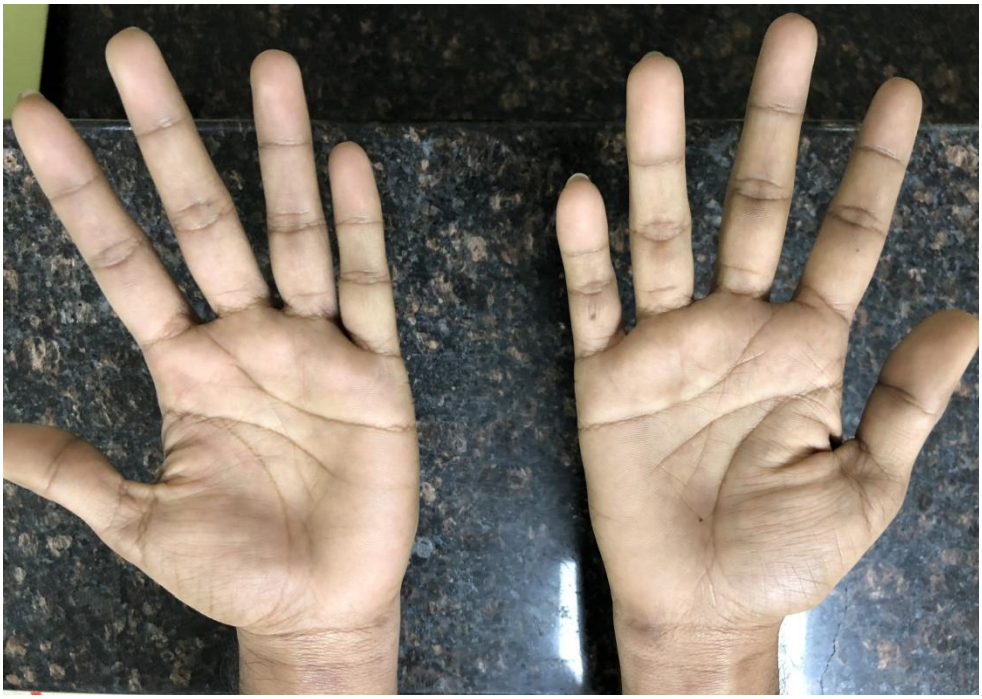


Figure 5: Image shows atrophy of thenar and hypothenar muscles of right hand,however the left thenar and hypothenar muscles show normal bulk.

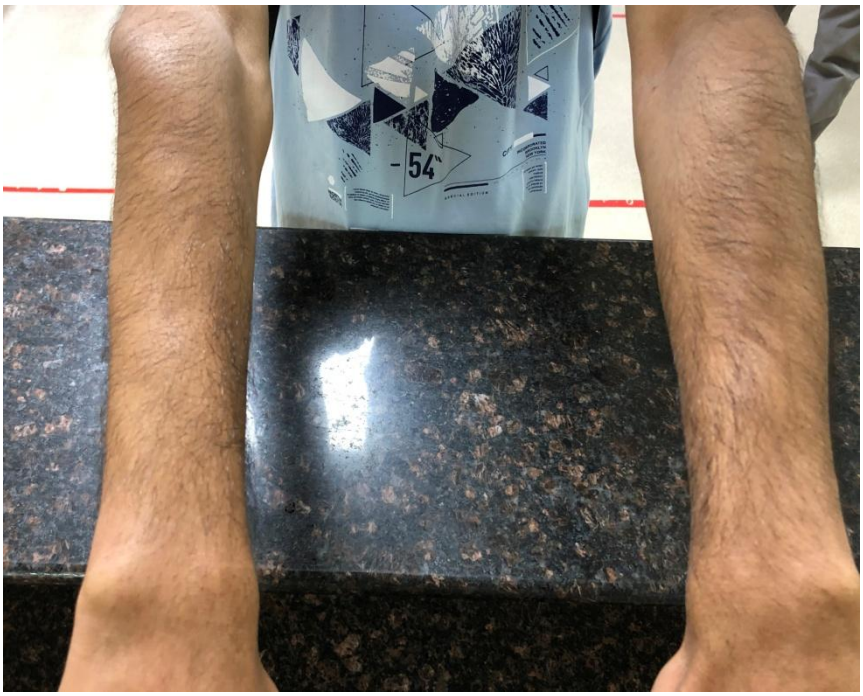
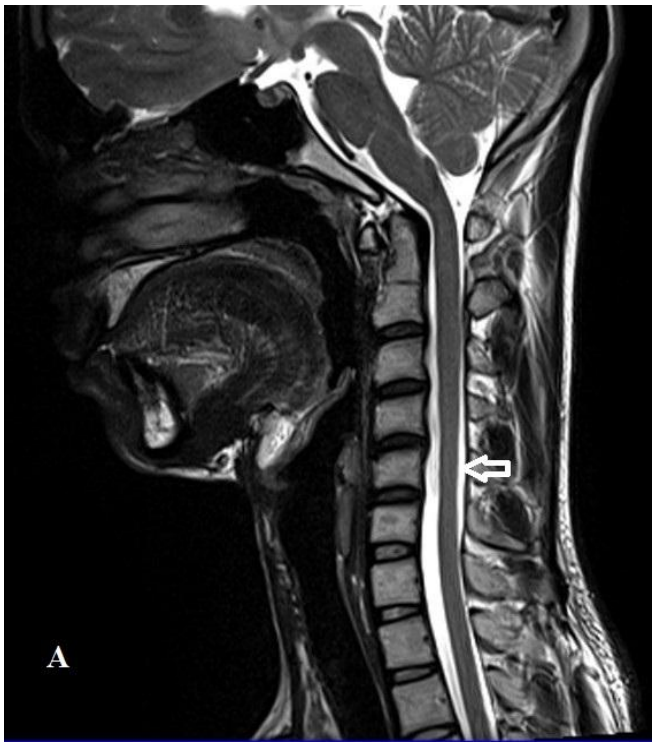


Figure 6: Image shows sparing of bilateral brachioradialis muscles.



CASE 2

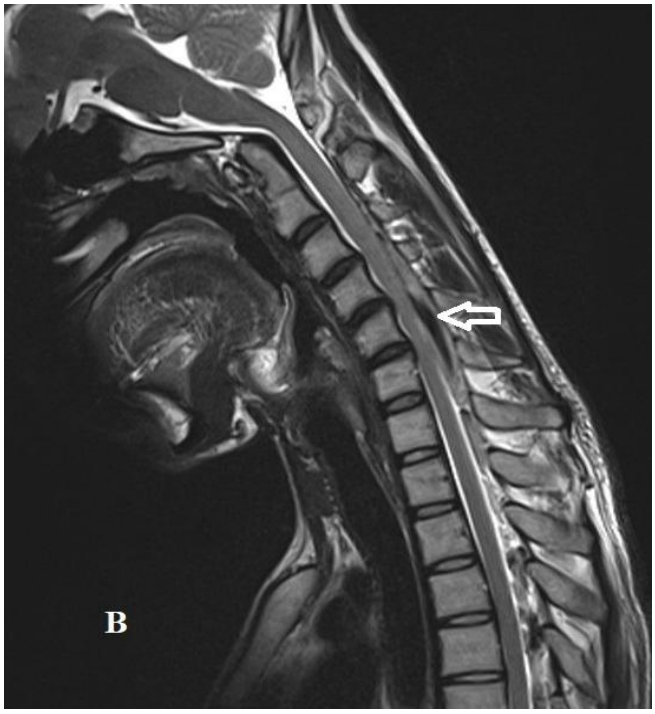


Figure 7 A,B: T2W Sagittal image in neutral position(A) demonstrates mild atrophy of cord with associated cord hyperintensity at C5,C6 level. T2W Sagittal image in flexion position(B) in same patient demonstrates detachement of posterior dural sac with multiple tortuous flow voids from C4 TO C6 level.(white arrow in B)

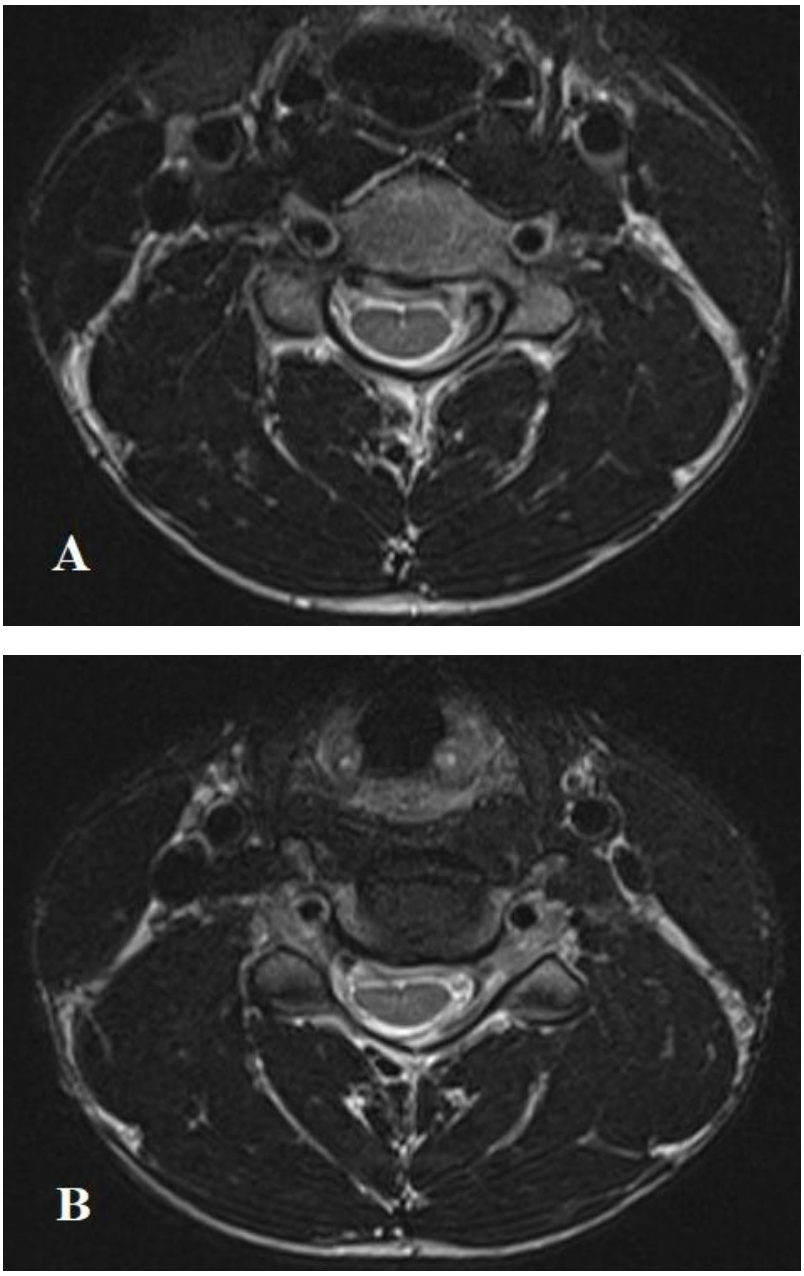
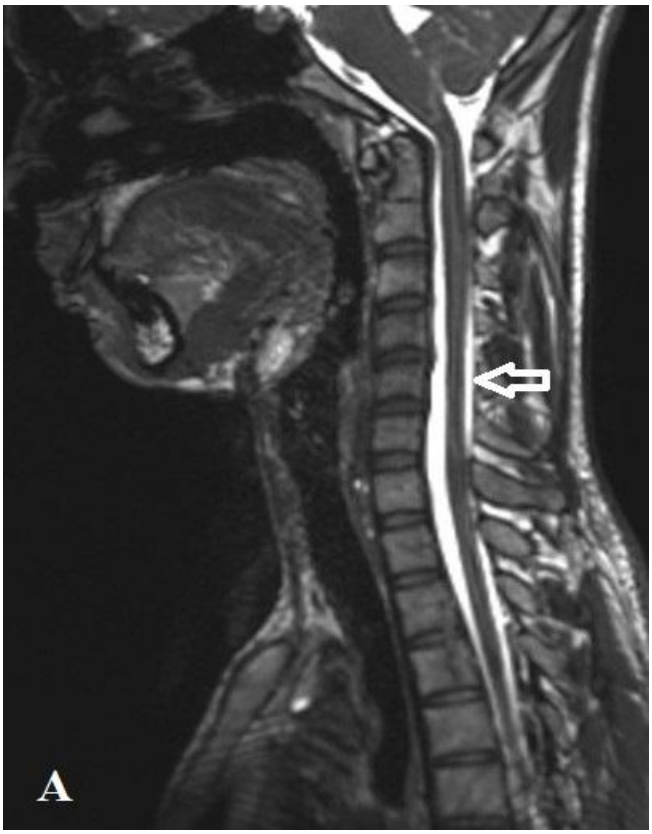


Figure 8 A,B: Axial T2W image at C3 level(A) demonstrates normal cord,there is flattening of cord with associated T2 hyperintensity at C5 level(B).



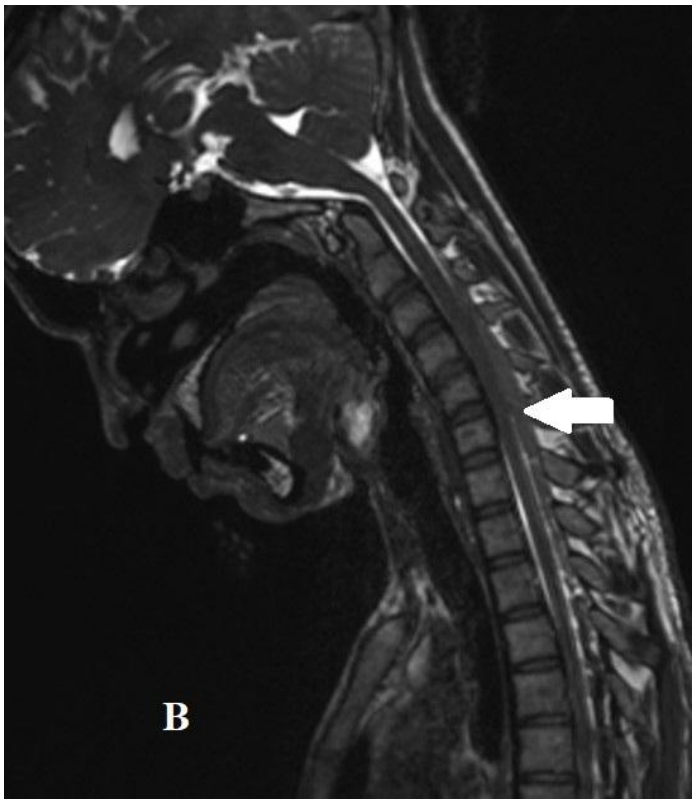


Figure 9 A,B: Sagittal **3D FIESTA** image in neutral position(A) demonstrates mild atrophy of cord (white arrow) at C5,C6 level. Sagittal **3D FIESTA** image in flexion position (B) in same patient demonstrates detachement of posterior dural sac with multiple tortuous flow voids extending from C4 to upper thoracic vertebral level.(bold white arrow in B)

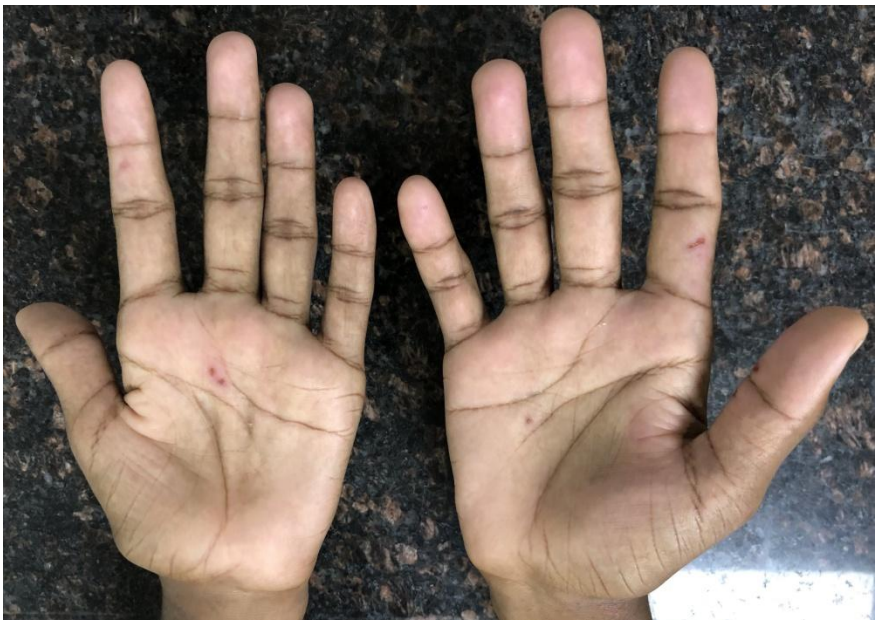


Figure 10: Image shows atrophy of thenar and hypothenar muscles of left hand,however the right thenar and hypothenar muscles show normal bulk.

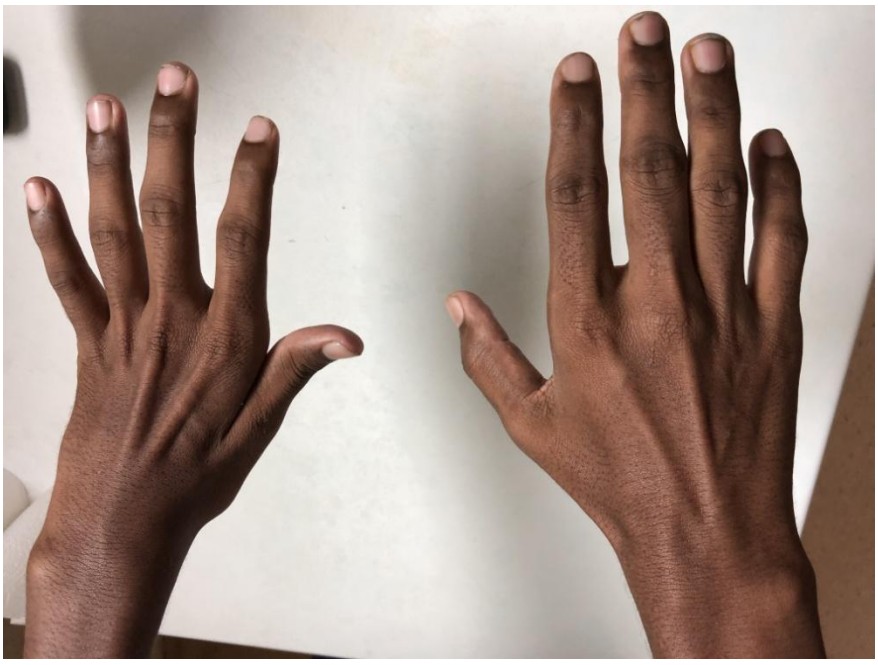
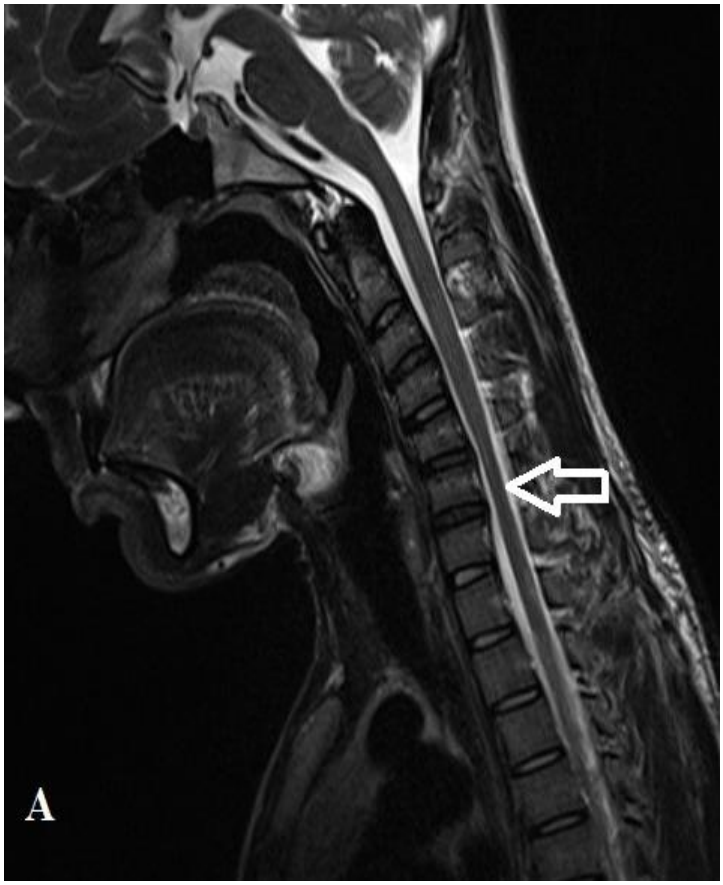


Figure 11: Image shows prominent extensor tendons with muscle atrophy of dorsal muscles of left hand,however the right hand is normal.

CASE 3



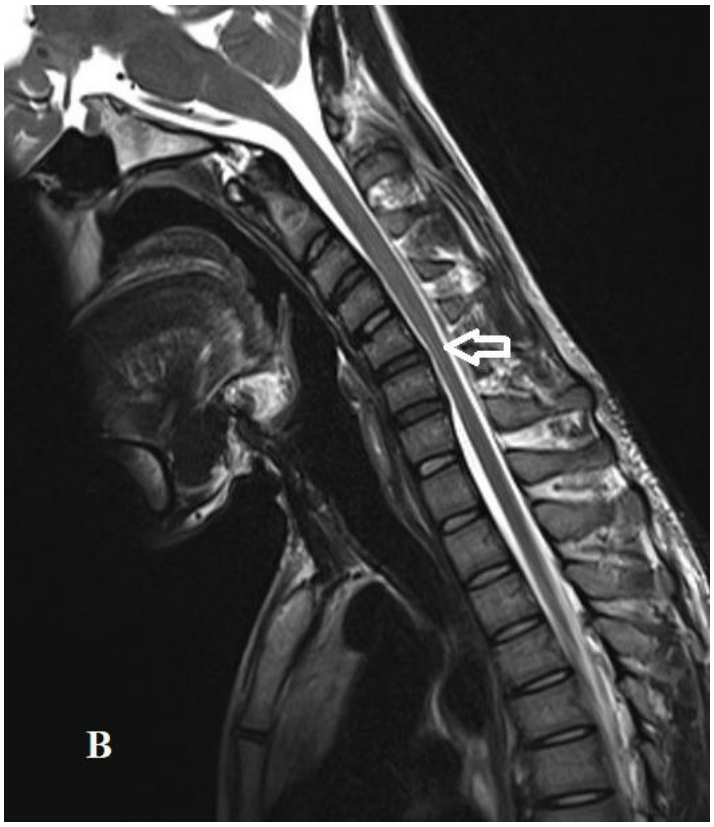


Figure 12 A,B: T2W Sagittal image in neutral position(A) demonstrates mild atrophy of cord with associated cord hyperintensity at C6,C7,T1 leve(white arrow). T2W Sagittal image in flexion position(B) in same patient shows the cervical spinal cord abutting the posterior border of cervical vertebrae from C4 to C6 level.(white arrow in B)

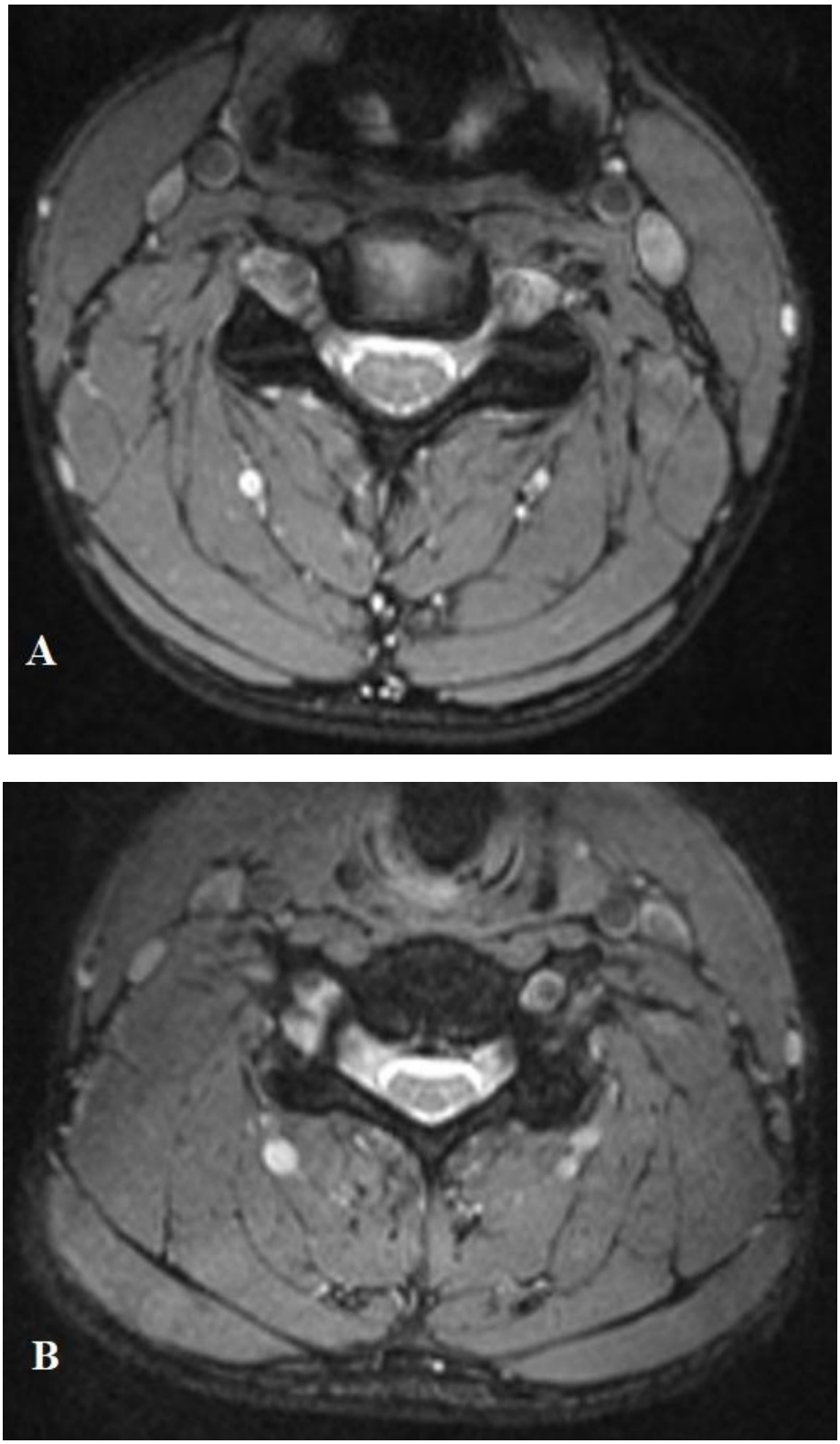


Figure 13 A,B: Axial GRE images at C3 level demonstrates normal cord(A),there is flattening of cord with associated T2 hyperintensity at C6 level(B).

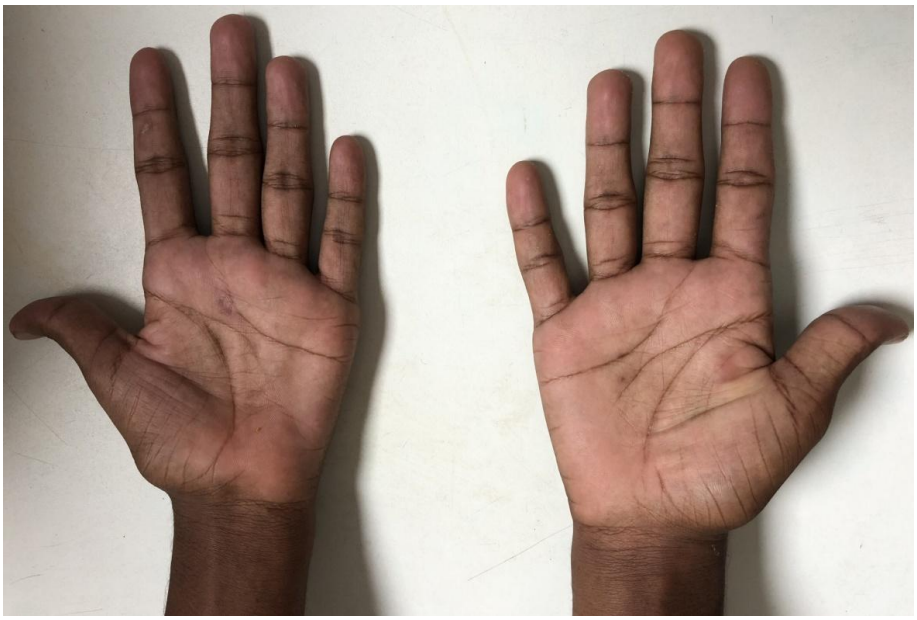


Figure 14 : Imageshows atrophy of thenar and hypothenar muscles of both hands(left more than right).

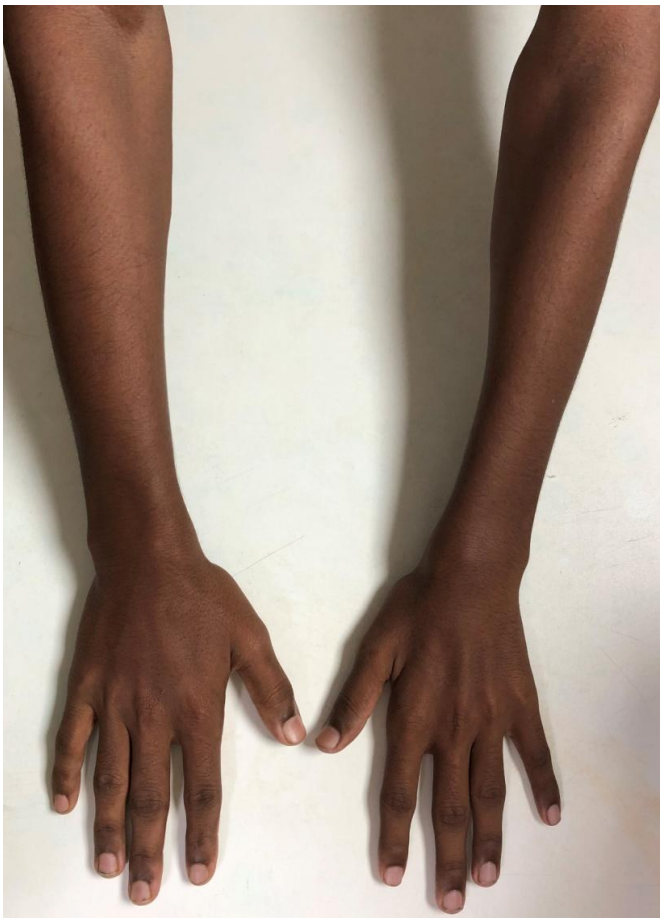


Figure 15: Image shows atrophy of distal muscles of left forearm,However the brachioradialis is spared in both upper limbs.

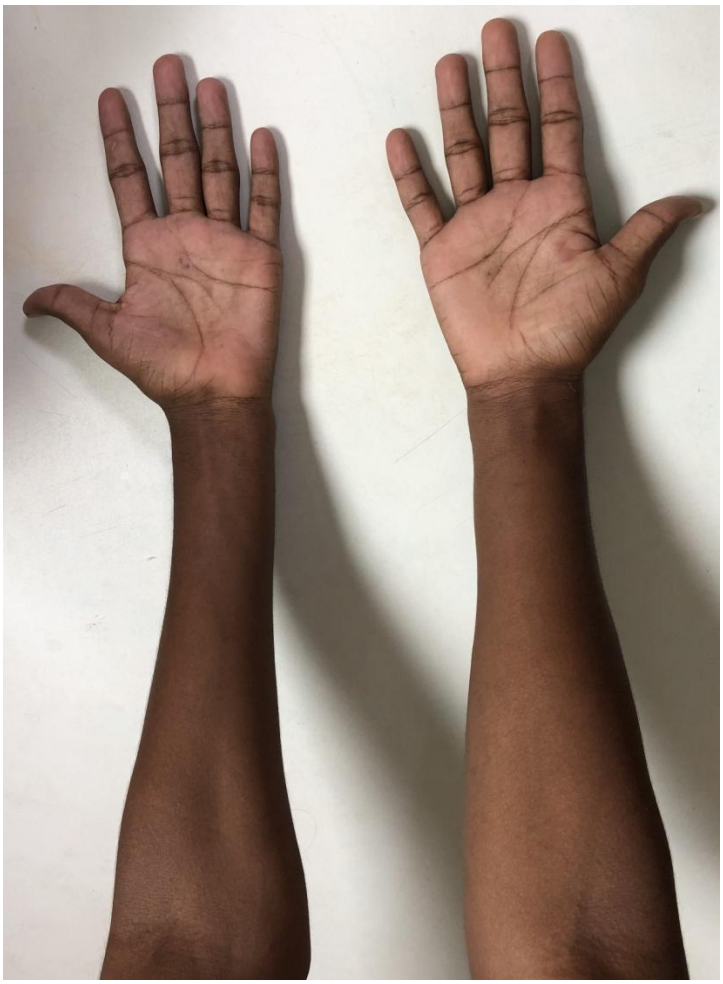
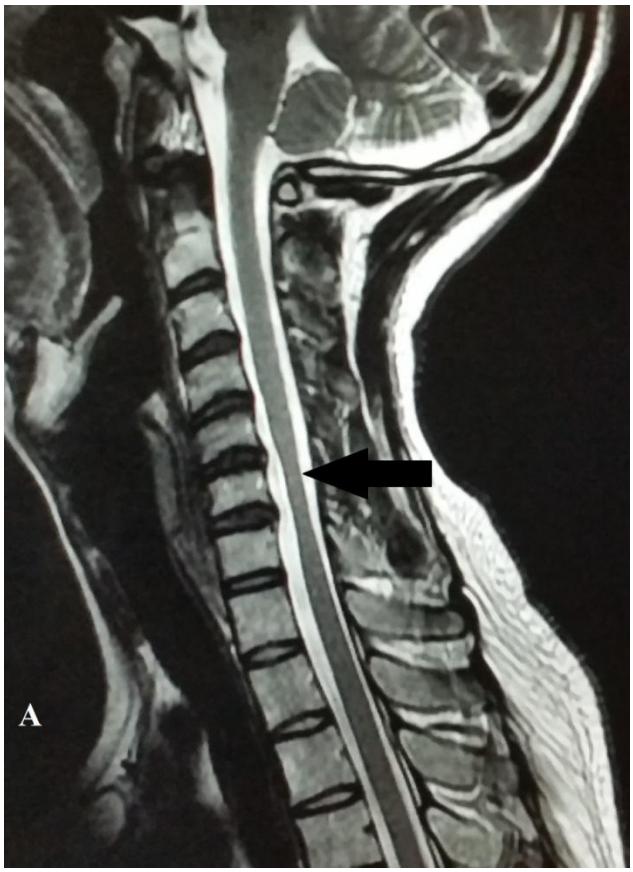


Figure 16: Image shows severe atrophy of flexor muscles of left forearm with sparing of right forearm.



CASE 4

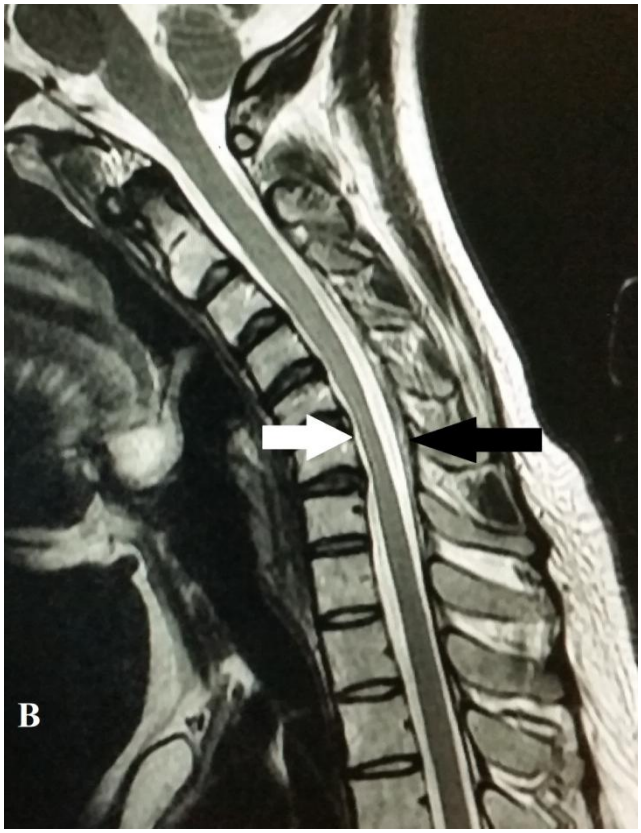
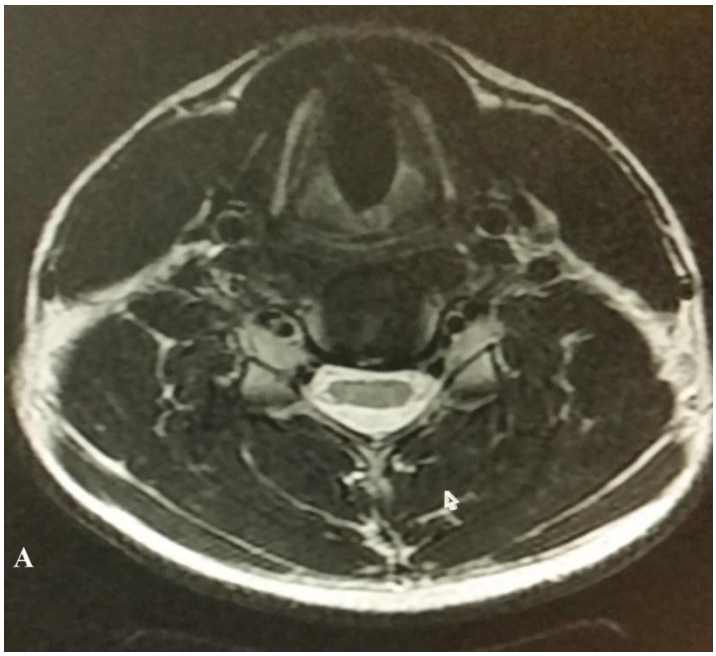


Figure 17 A,B: T2W Sagittal image in neutral position(A) demonstrates severe atrophy of cord with associated cord hyperintensity at C5,C6 level(black arrow. T2W Sagittal image in flexion position(B) in same patient shows the cervical spinal cord abutting the posterior border of cervical vertebrae from C4 to C7 level.(white arrow in B).There is mild detachement of posterior dural sac with few flow voids(black arrow in B)



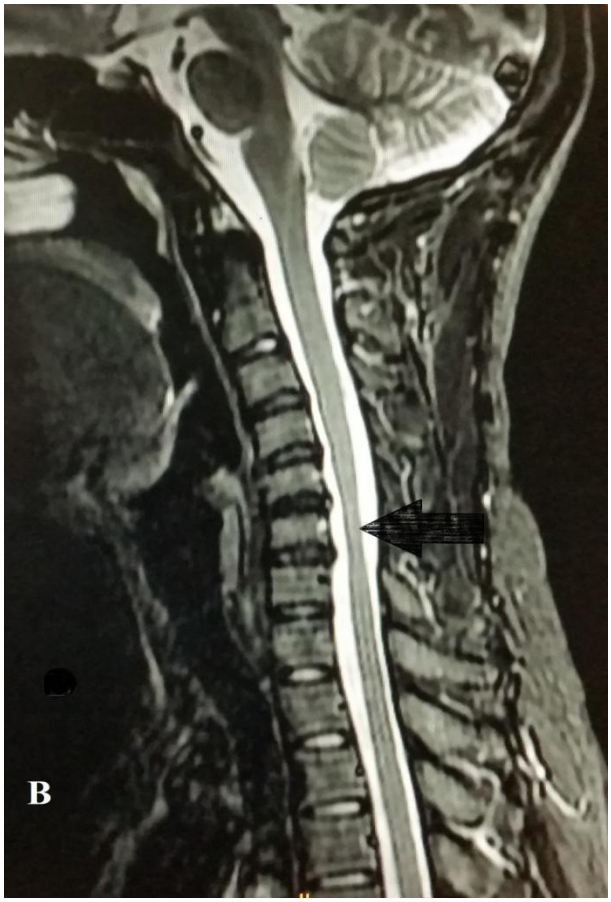
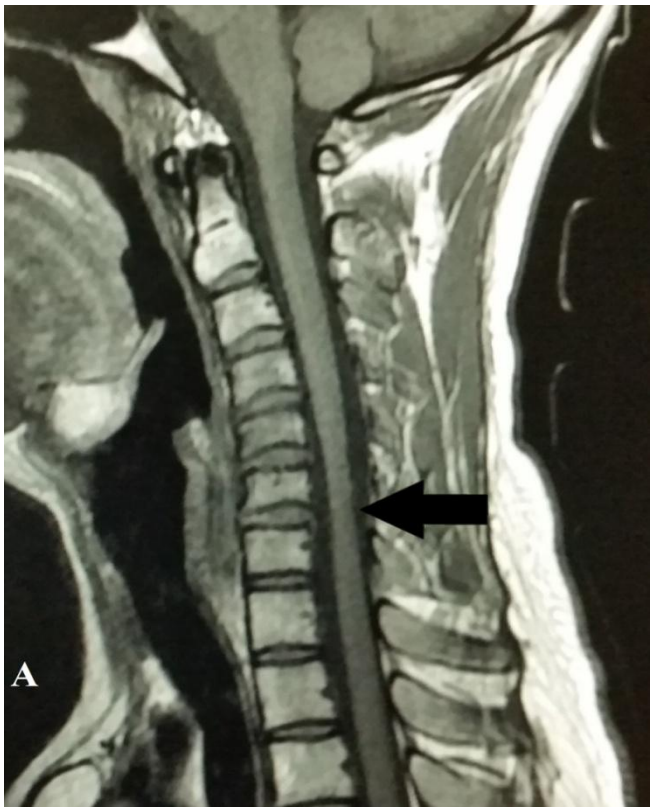


Figure 18 A,B: Axial T2W image(A) at C6 level shows flattening of cervical spinal cord with associated hyperintensity.Sagittal STIR image(B) shows severe atrophy of cord with associated cord hyperintensity at C5,C6 level(black arrow)



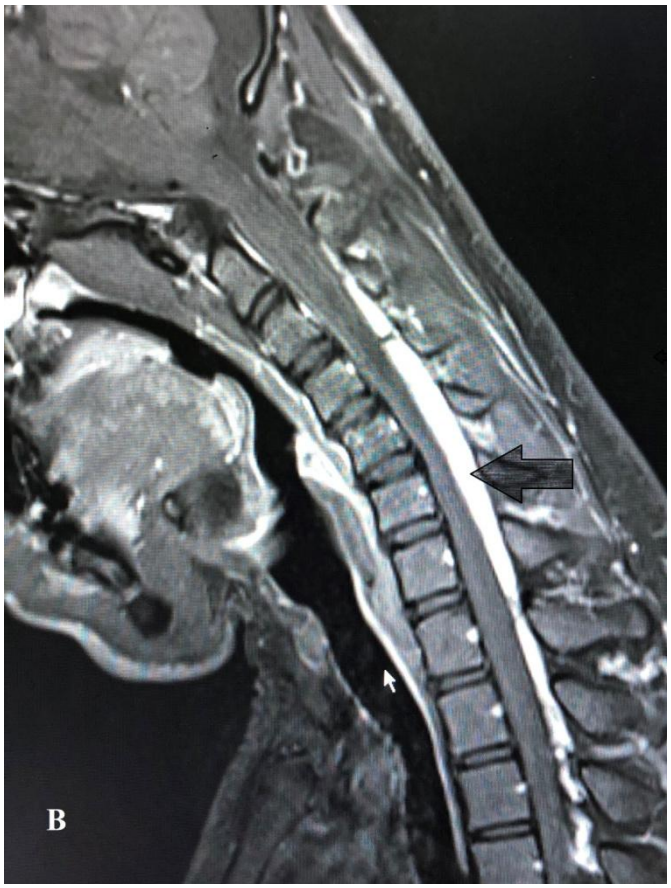


Figure 19 A,B: Sagittal T1W image in neutral position (A) shows severe atrophy of cord at C5,C6 level(bold black arrow). Sagittal T1 fat sat post contrast image in flexion position (B) shows intense enhancement (black arrow) of the posterior dural space extending from C3 vertebtal level upto the upper thoracic vertebrae.

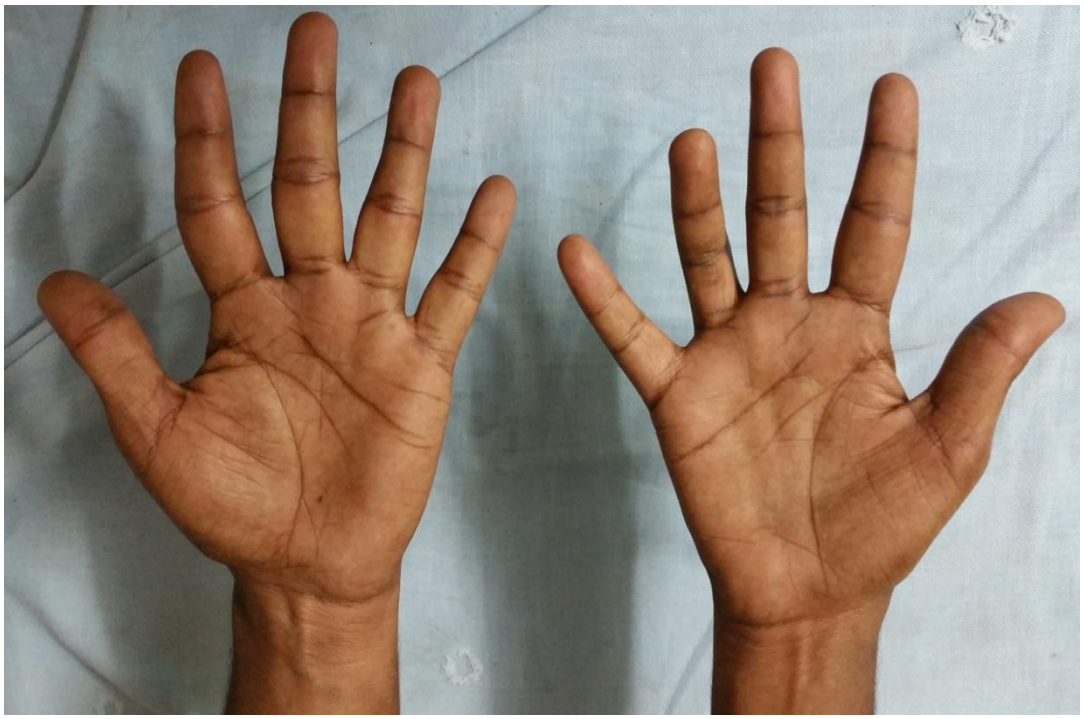


Figure 20: Image shows severe atrophy of thenar and hypothenar muscles of both hands.

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