Hirayama Disease: A Case of Invisible Compression

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CASE DESCRIPTION

- 39 year-old man with prior anterior discectomy and fusion of C4-7 in 2005, revision in 2017, posterior laminectomy and fusion from C2-T2 in 2018, presenting with neck pain and right hand numbness.
- Was scheduled for revision of fusion for progressive bilateral hand paresthesias and weakness but presented early due to acutely worsening neck and upper back pain after a fall.
- Imaging showed fused at C2-3, fused C3-7, but Right T1 screw broken and R T2 screw cap loose (Fig 1, 2).
- He was then discharged home and seen with neurosurgery as an outpatient. Ultimately, he underwent C2-T3 instrumentation and fusion with revision of C2-T1 bilateral decompression and revision of hardware.
- Of note, he had upper extremity weakness that began at age 8 and progressively worsened. Patient was without other neurologic or genetic conditions.

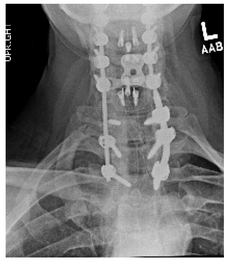

Fig 1. Sagittal view of right T1 screw broken and right T2 screw cap loose

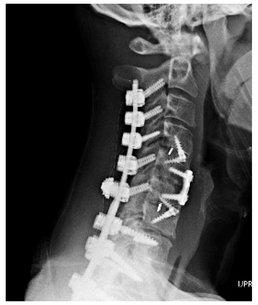

Fig 2. PA view of right T1 screw broken and right T2 screw cap loose

REHABILITATION COURSE

- Post revision and extension of fusion, patient was admitted to inpatient rehabilitation with significant right greater than left distal upper extremity weakness, specifically finger abduction and flexion, atrophy of hypothenar, thenar, and intrinsic hand muscles.
- Neurological testing of lower extremities was unremarkable. Sensation was grossly intact.
- He was placed in a Miami J collar, with Philadelphia collar for shower

Admission Level of Function:

Occupational Therapy:
ADLs: grooming min A with toothpaste, CG with standing at sink
Bed mobility supervision
Transfers supervision

Physical Therapy:
Bed mobility supervision
Transfers supervision
Ambulation 150 ft x 2 with no device, supervision; decreased cadence
Decreased endurance

Discharge Level of Function

Occupational Therapy
independent in feeding, oral hygiene, toilet hygiene, putting on and taking off shoes, set up assist for ubd

Physical Therapy
independent in transfers, ambulating 200ft on all surfaces independently, up and down flight or stairs with or without handrail

DISCUSSION

- Hirayama disease is a rare, often self-limiting, cervical myelopathy, predominant in Asian males.
- Other names: monomelic atrophy, distal juvenile muscular atrophy of the upper limbs
- During neck flexion, forward displacement of the posterior sac leads to spinal cord compression
- Tashiro et al proposed requirements for diagnosis:
  1. Mainly distal weakness and muscular atrophy affecting the forearm and hand
  2. Unilateral upper extremity involvement
  3. Age of onset 10-20 y/o
  4. Insidious onset with gradual progression, followed by stabilization after a few years
  5. No involvement of lower limbs
  6. No sensory disturbance, no reflex abnormalities
  7. Exclusion of other diseases (with imaging and EMG)

- Best evaluated by a T2 weighted cervical flexion MRI (Fig 3.)
  - Findings during neck flexion:
    - asymmetric medullary flattening
    - prominence of the posterior epidural venous plexus
    - anterior movement of the posterior dural sac
- Only EMG abnormalities may be:
  - decreased magnitude of compound motor action potential in affected muscles
- Management:
  - Cervical collar that blocks the flexion of the neck
  - Muscle strengthening exercises for hands
  - Vertebral fusion surgery for advanced cases
- For this patient, after review of history and prior imaging, neurosurgery suspected Hirayama disease.
  - No other diagnosis explained the early onset weakness and worsening myelopathic changes in areas well decompressed.
  - Unfortunately, diagnosis could not be confirmed due to inability of patient to remain flexed during MRI.

CONCLUSIONS

- Literature describing Hirayama disease is limited
- HD should be considered in patients presenting with history of weakness and atrophy of upper limbs and hands, with insidious onset and slowly progressive course
- HD is diagnosed with cervical flexion MRI
- Patients should be recognized early and placed in a cervical collar
- Rehabilitation focuses on strengthening of the distal musculature, coordination, and avoidance of flexion-based exercises until resolution

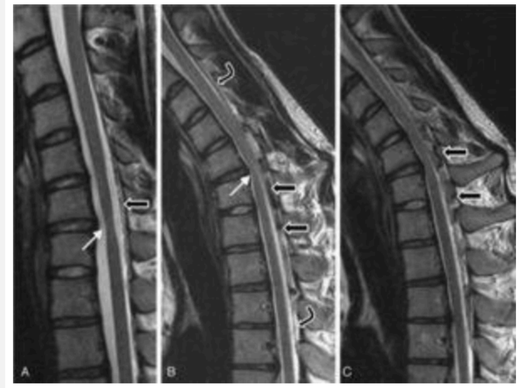

Fig 3. Flexion Based MRI from Agundez et al in a patient with Hirayama Disease.

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