Case report: A 32-Year-Old Man With Painless Bilateral Shoulder Girdle Weakness and Atrophy

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HISTORY & EXAMINATION

•A 32-year-old man, progressive weakness and wasting of left shoulder girdle muscles for 14 months, and similar symptoms on the right side for 4 months.
•No neck pain, headache, vomiting, or fever.
•2 years back, h/o fall from bike, lacerations over the face, chin, and trunk, managed conservatively, and recovered without neurologic deficit.
•10 months later, he had left shoulder weakness.
•Examination revealed gynecomastia and purple striae over his lower abdomen. He had wasting of the bilateral supraspinatus, infraspinatus, deltoid, biceps, triceps, and pectoralis muscles, more marked on left side .
•Muscle power MRC grade 2 in the left deltoid, pectoralis, supraspinatus, and infraspinatus; grade 3 in the left biceps and triceps, right deltoid, pectoralis, supraspinatus and infraspinatus; and grade 4 in the right biceps and triceps.
•Upper limb deep tendon reflexes were reduced.
•The lower limb examination was normal.
• Touch, pain, and temperature sensation was reduced over the outer aspects of his left shoulder, arm, forearm, and hand.
• Sensation on the right side was normal.

ANALYSIS

•Patient presented with painless, progressive, asymmetric (left > right) proximal weakness of both upper limbs with sensory impairment in the left C5-C8 dermatomes.
•The weakness and atrophy suggested lower motor neuron type of involvement, possibly due to lesions of the anterior horn cells (AHCs), nerve roots, brachial plexus, peripheral nerves, neuromuscular junction, or muscles.

INVESTIGATION

•CBC & biochemistry were normal.
•NCS bilateral median, ulnar, peroneal, and sural nerves were normal.
•Concentric needle EMGof the bilateral supraspinatus, deltoid, biceps, triceps, left extensor digitorum communis, first dorsal interossei, and abductor pollicis brevis revealed fibrillations, positive sharp waves and long duration, high amplitude polyphasic motor unit potentials with reduced interference, but was normal in the lower limb muscles.
• Edx findings suggested either motor root or AHC involvement of C5-C8 because of the normal sensory conduction findings.
•His median and ulnar CMAP & SNAP normal. Morning cortisol (239.6 nmol/L) and thyroid stimulating hormone (4.96 mIU/L) levels were normal, but prolactin was elevated (521 mIU/L; normal 85–319 mIU/L).
•Craniocervical MRI downward displacement of cerebellar tonsil - brain sagging.
•Dural enhancement of the posterior fossa and cervical spine and CSF leakage posteriorly at the C1-C2 level .
•Lumbar CSF opening pressure was 6cm H2O, and CSF analysis normal.
• CT myelography revealed contrast extravasation along the bilateral perineural space and paravertebral soft tissue at the C1-C2 level

IMAGES

(A)Clinical photograph showing wasting of both deltoids, pectoralis, and biceps muscles. He has gynecomastia. (B) MRI of the cervical spine and craniovertebral junction shows downward displacement of the tonsil on T1 sagittal section (arrow), (C) patchy pachymeningeal enhancement (arrow), and (D) CSF leakage at C2 level (arrow). (E) Gadolinium-enhanced cranial MRI shows pachymeningeal enhancement. (F) CT myelography shows leakage of CSF at the C1-C2 level (arrow), and there is (G) seepage into the paravertebral area posteriorly. (H and I) Postoperative CT myelography shows no leakage of CSF.

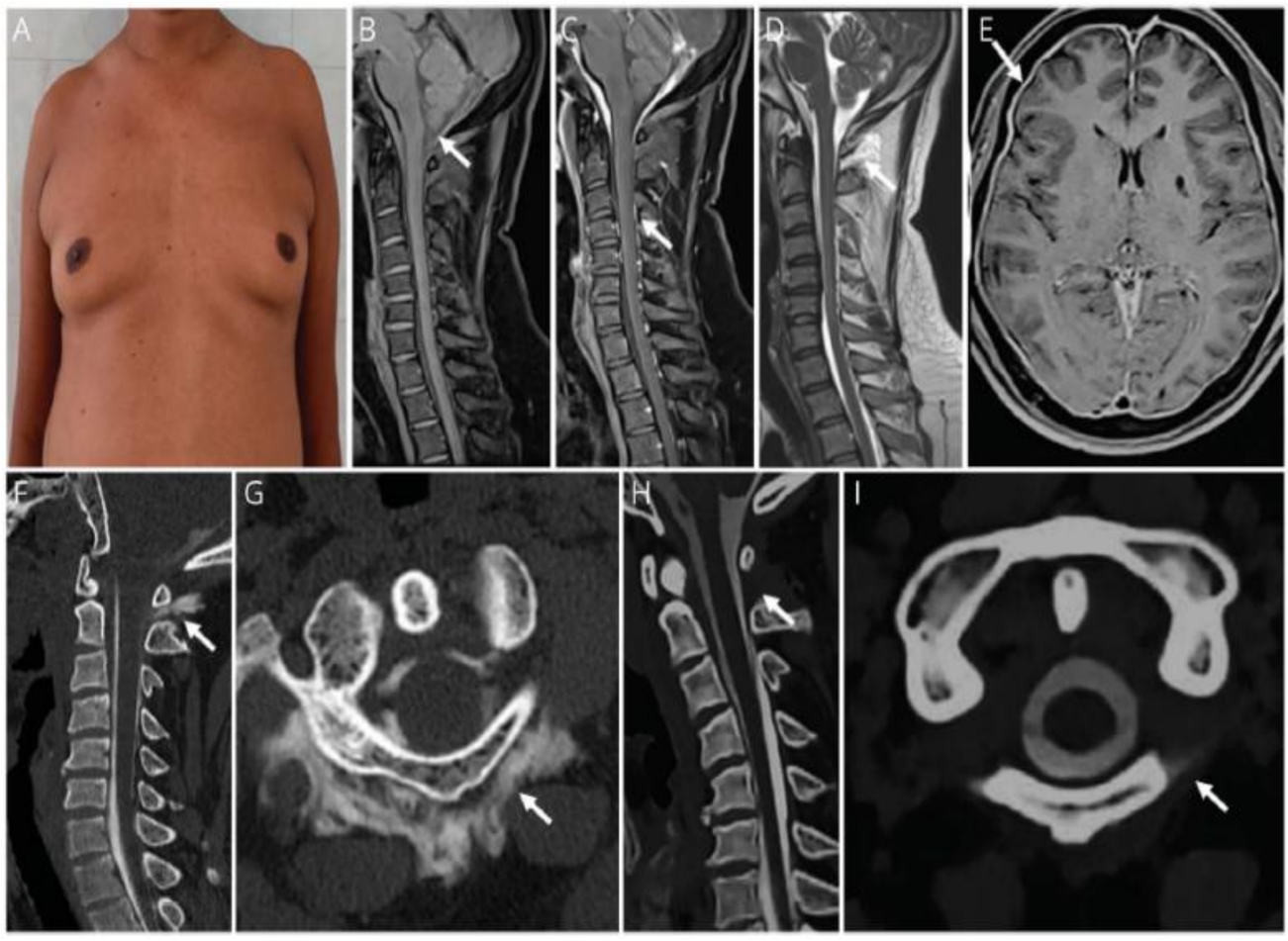
MANAGEMENT

•Advised to take fluid and caffeine.
•An epidural blood patch (EBP) is a standard procedure for spinal CSF leaks, was attempted , but terminated because of seepage of dye from the epidural space to the intrathecal space, which can cause chemical meningitis and compressive myelopathy.
•Subsequently underwent neurosurgical repair of the dural tear, which is indicated in patients with large or multiple leaks, and who fail to respond to a blood patch.
• A 1 × 1 cm dural tear was noted dorsally at C1-C2, confirmed using fluorescein dye.
•The tear was repaired using a muscle patch and reinforced using duragen.
•A repeat CT myelogram did not reveal leakage .
• At 6-month follow-up, weakness and wasting had stabilized.
•Mild improvement in the bulk of left supraspinatus and infraspinatus.
•Repeat serum prolactin level was normal (211mIU/L), although gynecomastia persisted.

CLINICAL DIFFERENTIAL DIAGNOSIS

Table Clinical Differential Diagnosis of Bilateral Brachial Amyotrophy

Localization	Supporting features	Undermining features	Specific diagnoses
Muscle	➢ Upper limb girdle pattern of weakness and wasting ➢Pectoral atrophy causing exaggerated anterior axillary crease	➢Sensory involvement ➢Absence of facial muscle weakness ➢Absence of Beevor sign ➢No scapular winging	➢Facioscapulohumeral muscular dystrophy ➢LGMD
Neuromuscular junction	➢Proximal weakness	➢No fluctuations ➢No ocular/bulbar/facial muscle involvement ➢Muscle wasting and sensory findings	➢Myasthenia gravis
Peripheral nerves	➢Sensory deficit and muscle weakness and wasting	➢Only proximal pattern of weakness	➢Multifocal motor neuropathy
Brachial plexus	➢Bibrachial neurogenic amyotrophy ➢Sensory deficit ➢Asymmetric ➢Triggered by trauma	➢Bilaterality is rare ➢Absence of pain ➢Progressive course	➢Brachial neuritis
Roots	➢Sensory motor weakness ➢Bilateral proximal weakness ➢Significant atrophy ➢Radicular distribution of sensory loss	➢Absence of radiating pain	➢Cervical spondylosis with compressive radiculopathy
Anterior Horn cell	➢Bibrachial weakness ➢Depressed tendon reflex in the upper limb ➢Gynecomastia ➢Male sex	➢Sensory loss ➢Absence of bulbar symptoms ➢No infertility	➢Kennedy syndrome ➢Amyotrophic lateral sclerosis
Intramedullary central cervical spinal cord lesion	➢Upper limb flaccid weakness and wasting ➢Sensory loss in dermatomal pattern ➢History of accidental injury	➢Absence of typical pattern of suspended and dissociated sensory loss ➢Asymmetric weakness ➢Absence of lower limb symptoms	➢Syringomyelia



BIBRACHIAL AMYOTROPHY SECONDARY TO CSF LEAK