Rare case of syringomyelia(lipomyelocele) presented with Paraplegia in post-partum female

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INTRODUCTION

Syringomyelia, at its core, is a disorder of abnormal cerebrospinal fluid (CSF) circulation. A syrinx is a fluid-filled cavity thatanatomically lies within the spinal cord parenchyma or the central canal. This entity is most frequently associated with a CM-I[1],although other known causes include spinal cord tumor, trauma, and post-traumatic or infectious adhesive arachnoiditis.Although syringomyelia predominantly presents with sensory symptoms such as pain and temperature
insensitivity, in most cases it is an incidental finding.The discovery of syringomyelia is becoming more common because of theincreased use of MRI in the routine evaluation of back and neck pain[2]

The natural history of patients with syringomyelia is variable and unpredictable punctuated with periods of stability and progression. Although professionals do not completely understand the natural history of syringomyelia, the clinical course progresses over months to years, with an early rapid deterioration that gradually slows down[3]. It is also understood that sudden jolting of the head, a prolonged bout of coughing may bring a sudden onset of symptoms in a previously asymptomaticpatient presumably because of an increase in tonsillar descent.
Syringomyelia accounts for up to 5% of paraplegia.The quality of life in syringomyelia patients is comparable with that ofpatients with heart failure or malignant neoplasms.

Case report:

A 29 years old female patient presented with bilateral lower limb weakness and inability to walk even with support and not able to bearing weight. Not able to bend knee. Associated with lower back pain. Patient also has complaint of constipation and urinary incontinence since 3 month.
Past history of:
During 2nd delivery child was dead after 8th day of delivery and patient bedridden for 1 month. During this time 3rd delivery pt was bedridden for 3 month after delivery.
On examination: on thoracic spine small dipping since childhood.Power in upper both limb normal but both lower limb zero.
Tone increase in both lower limb.both planter are extensor.all reflexes are exaggerated in bilateral lower limb. On investigation:all blood reports are normal.

Diagnosis:

MRI

MRI with and without contrast is the investigation of choice. It delineates relevant anatomy and allows accurate visualization of the syrinx in both sagittal and axial planes. MRI easily reveals the location, size, and extent of the syrinx cavity, the degree of cerebellar tonsillar ectopia. A ubiquitous feature in patients with associated CM1 is compression of retro-cerebellar CSF spaces[4]. MRI also helps to rule out cystic lesions or spinal tumors. Leptomeningeal enhancement indicates infection. MRI can also reveal any arachnoid scarring. One can also study syrinx progression over months or years to document the natural historyof syringomyelia.

Dynamic MRI or Cardiac Gated CINE-MRI Flow Study

These can analyze CSF hydrodynamics non-invasively. It can diagnose CSF velocity/flow disturbance at the foramen magnum (esp. in patients with <5mm tonsillar ectopia), visualize spinal cord wall motion, and syrinx fluid motion at rest during the cardiac systole and diastole.It is also useful to document postoperative CSF flow changes and objective improvements.

Myelography with High-Resolution CT Scan
This scan is indicated where MRI cannot be used (patients with metallic implants in the joints, cardiac pacemaker). Delayed CT scans can visualize dye leached into the syrinx cavity. However, some authors have criticized CT myelography for having a lowsensitivity in detecting CSF blockage.

Electromyography has no diagnostic value in syringomyelia, but it helps to rule out peripheral neuropathy causing paresthesias.Treatment:C7-D1,D2 laminectomy +excision of space occupying lesion with duroplasty.

Discussion:
The basic pathology in syringomyelia is a progressively expanding cavity in the central spinal canal. Increased pulse pressure in subarachnoid spaces forces CSF through spinal cord into the syrinx[5]. This expanding CSF filled "syrinx" compresses the spinothalamic tract neurons decussating in the anterior white commissure. However, the posterior columns are spared as they are located distally. This results in loss of pain and temperature sensation with preserved touch and vibratory sense (segmental dissociated sensory loss).

Differential Diagnosis
Spinal intramedullary tumors such as hemangioblastoma, ependymoma, gliomas: Tumors may secrete exudative fluid (high in protein content) causing micro-cysts that can eventually coalesce. Most of these intramedullary tumors will enhance on contrast MRI (a syrinx does not enhance on contrast MRI). However, a true syrinx can occur within a tumor)
Spinal intramedullary cystsMyelomalacia
Arachnoid cysts
Glio-ependymal cysts
Residual central spinal canal: The central canal of the spinal cord involutes with age. A persistent central canal is NOT an anomaly.

Prognosis
Professionals still do not understand the natural history of syringomyelia; it is unpredictable and highly variable which makes its prognostication very difficult. Although prognosis depends on the etiology, the degree of neurological deficit and the site and size ofthe syrinx cavity, syrinx diameter of more than 5 mm, and associated edema predict a rapid deterioration. The rarity of the condition, variable natural history, and short follow-up make treatment results assessment difficult.However, early surgery minimizesdeficits and has better outcomes.
Syringomyelia is chronic progressive condition and syrinx appears to be the source of symptoms. Appropriate diagnosis and treatment of the pathology is necessary so that expansion of syrinx cavity is reduced and quality of life and functionality of patientsis maintained .[6]

Complications
Myelopathy is the major complication because of the disease process itself. This can further lead to spasticity which may progressto paraplegia/quadriplegia, decubitus ulcers, recurrent pneumonia, and bowel and bladder dysfunction.

Neurological complications following surgery include cerebrospinal fluid (CSF) leaks, infection, hemorrhage, recurrence of the syrinx

Conclusion:
We report case of 29 years old patient present with paraplegia with exaggerated lower limb reflexes.on examination dipping on thoracic vertebra and confirm by MRI spine that there is presence of syrinx formation of peripheral aspect of cord extending from C7 -D1 to D2-3 level and nueral element herniate through this syrinx with multiple fat intensity within the spinal canal suggestive oflipomyelocele There are various concepts for understanding pathophysiology of Syringomyelia like cerebral piston theory, intramedullary shunt theory,and increased spinal subarachnoid pressure. Most patients are on conservative management while decompression like laminectomy, lysis of adhesion and craniocervical decompression is considered for pts with progression of symptoms.[7]
If syrinx is the result of post traumatic or post arachnoidictic syringomyelia, the treatment is best directed to disabling the likely filling mechanisms by opening up the CSF pathways and leaving Dura open to create an artificial meningocoele.[8]

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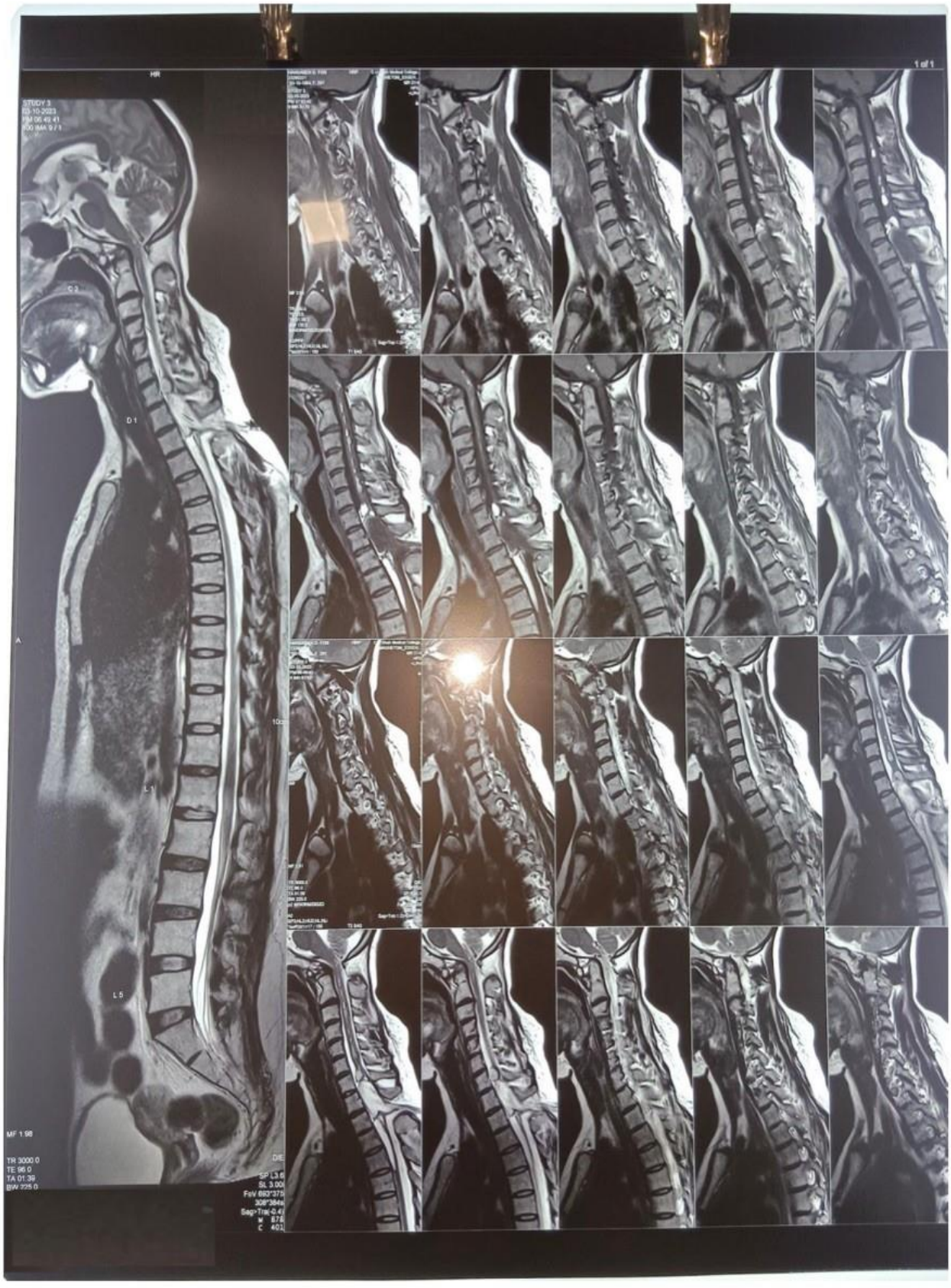
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HR
STUDY 3
C2
D1
L1
L5
TR 3000.0
TE 96.0
TA 01:39
BW 225.0