Case Report

A rare case of Syringomyelia with Arnold – Chiari Malformation Type 1 - A case report

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Abstract

Syringomyelia is a pathologic cystic cavity within the spinal cord containing cerebrospinal fluid (CSF). It is commonly seen as a complication of an Arnold-Chiari type 1 malformation, which is the herniation of cerebellar tonsils through foramen magnum into cervical spinal canal. Syringomyelia can also occur as complication of hemorrhage, tumor, meningitis, arachnoiditis, or trauma. Symptoms usually begin to appear in early and middle life. These symptoms usually consist of pain, dissociated sensory loss and weakness that present and progress gradually. We herewith report a rare case of syringomyelia and associated Chiari I malformation presenting with dissociated sensory impairment in neck region with headache and neck pain Treatment in these cases is surgical decompression. Recovery with significant decrease in sensory loss and relief in headache and neck pain.

Key words

Syringomyelia, Arnold –Chiari malformation Type 1, Headache, Neck pain, Dissociated sensory loss.

Introduction

Syringomyelia is the development of a fluid-filled cavity or syrinx within the spinal cord. Hydromyelia is a dilatation of the central canal by cerebrospinal fluid (CSF) and may be included within the definition of syringomyelia. The following are types of syringomyelia.

Syringomyelia with fourth ventricle communication

About 10% of syringomyelia cases are of this type. This communication can be observed on

MRI. In some cases, a blockage of CSF circulation occurs. A shunt operation may be the best therapeutic option for these patients.

Syringomyelia due to blockage of CSF circulation (without fourth ventricular communication)

Representing at least 50% of all cases, this is the most common type of syringomyelia. Obstruction of CSF circulation from the basal posterior fossa to the caudal space may cause syringomyelia of this type. The most common example is Arnold-Chiari malformation, which is also associated with communicating syringomyelia. Other causes include the following: 1.Basal arachnoiditis (postinfectious, inflammatory, postirradiation, blood in subarachnoid space) [1], 2. Basilar impression or invagination, 3. Meningeal carcinomatosis and 4. Pathological masses (arachnoid cysts, rheumatoid arthritis pannus, occipital encephalocele, tumors) [2].

Syringomyelia due to spinal cord injury

Fewer than 10% of syringomyelia cases are of this type. Mechanisms of injury include (1) spinal trauma, (2) radiation necrosis, (3) hemorrhage from aneurysm rupture or arteriovenous malformation [3],or in a tumor bed, (4) infection (spinal abscess, human immunodeficiency virus, transverse myelitis), and (5) cavitation following ischemic injury or degenerative disease.

Syringomyelia and spinal dysraphism

Spinal dysraphism may cause syringomyelia through a variety of mechanisms, including those mentioned under the previous three categories. Identification and treatment of associated dysraphism has the greatest impact on arresting progression of syringomyelia.

Syringomyelia due to intramedullary tumors

Fluid accumulation is usually caused by secretion from neoplastic cells or hemorrhage. The tumors most often associated with syringomyelia are ependymoma and hemangioblastoma. Extramedullary intradural and extradural tumors are considered separately under the second category because the mechanism of syrinx formation is blockage of the CSF pathway.

Idiopathic syringomyelia

Idiopathic syringomyelia has an unknown cause and cannot be classified under any of the previous categories [4]. Surgical decompression can help in some patients with remarkable neurologic deficit.

Pathophysiology

The exact pathogenesis is still unknown. Frequently cited theories are those of Gardner, William, and Oldfield [5, 6, 7].

Etiology

Etiology of syringomyelia often is associated with craniovertebral junction abnormalities.

Bony abnormalities include 1. Small posterior fossa, 2. Platybasia and basilar invagination, 3. Assimilation of the atlas, 4. Soft-tissue masses of abnormal nature include the following: 5. Tumors (e.g. meningioma at foramen magnum), 6. Inflammatory masses

Neural tissue abnormalities include, 1. Cerebellar tonsils and vermis herniation, 2. Chiari malformation.

Membranous abnormalities include 1. Arachnoid cysts [8], rhombic roof, or vascularized membranes, 2. Post-hemorrhagic or post-inflammatory membranes.

Other etiologies not associated with craniovertebral abnormalities may include,

1. Arachnoid scarring related to spinal trauma, 2. Arachnoid scarring related to meningeal inflammation, 3. Arachnoid scarring related to surgical trauma, 4. Subarachnoid space stenosis due to spinal neoplasm or vascular malformation, 5. Subarachnoid space stenosis, with possible scarring, related to disk and osteophytic disease and 6. Idiopathic.

Epidemiology

Estimated prevalence of the disease is about 8.4 cases per 100,000 people. It is more common in men than in women. The disease appears in third or fourth decade of life. Syringomyelia may develop rarely in pediatric or geriatric ages [9].

Prognosis

Prognosis depends on the cause, the neurological dysfunction, and the location of the syrinx. Patients presenting with moderate or severe neurological deficits fare much worse than those with mild deficits. Patients with central cord syndrome have poor response to treatment.

Natural history of syringomyelia still is not well understood. Although older studies had suggested that 20% of patients died at an average age of 47 years, mortality rates are likely lower in current situation as a result of surgical interventions and better treatment of complications associated with significant paresis, such as pulmonary embolism [10].

Complications - Myelopathy is the most serious consequence of syringomyelia.

The following are the seven grade classifications of disability from myelopathy according to the **Modified Nurick Classification**.

• Grade 0 - No root signs or symptoms
• Grade I - Root signs or symptoms; no evidence of cord involvement
• Grade II - Signs of cord involvement; normal gait
• Grade III - Mild gait abnormality; able to be employed
• Grade IV - Gait abnormality prevents employment
• Grade V - Able to ambulate only with assistance
• Grade VI - Chairbound or bedridden

Complications due to myelopathy include 1. Recurrent pneumonia, 2. Paraplegia or quadriplegia, 3. Decubitus ulcers, and 4. Bowel and urinary dysfunction.

Mortality/morbidity

Assessing treatment results is difficult because of the rarity of syringomyelia, variability of presentation and natural history, and the relatively short follow-up in most studies.

Syringomyelia Clinical Presentation

Sensory

Syrinx interrupts the decussating spinothalamic fibers that mediate pain and temperature sensibility, resulting in loss of these sensations, while light touch, vibration, and position senses are preserved (dissociated sensory loss). When the cavity enlarges to involve the posterior columns, position and vibration senses in the feet are lost; astereognosis may be noted in the hands. Pain and temperature sensation may be impaired in either or both arms, or in a shawl like distribution across the shoulders and upper torso anteriorly and posteriorly. Dysesthetic pain, a common complaint in syringomyelia, usually involves the neck and shoulders, but may follow a radicular distribution in the arms or trunk. The discomfort, which is sometimes experienced early in the course of the disease, generally is deep and aching and can be severe.

Motor

Syrinx extension into the anterior horns of the spinal cord damages motor neurons (lower motor neuron) and causes diffuse muscle atrophy that begins in the hands and progresses proximally to include the forearms and shoulder girdles. Claw hand may develop. Respiratory insufficiency, which usually is related to changes in position, may occur.

Autonomic

Impaired bowel and bladder functions usually occur as a late manifestation. Sexual dysfunction may develop in long-standing cases. Horner syndrome may appear, reflecting damage to the sympathetic neurons in the intermediolateral cell column.

Extension of the syrinx

A syrinx may extend into the medulla, producing a syringobulbia [11, 12]. This syndrome is characterized by dysphagia, nystagmus,

pharyngeal and palatal weakness, asymmetric weakness and atrophy of the tongue, and sensory loss involving primarily pain and temperature senses in the distribution of the trigeminal nerve. Rarely, the syrinx cavity can extend beyond the medulla in the brain stem into the centrum semiovale (syringocephalus). Lumbar syringomyelia can occur and is characterized by atrophy of the proximal and distal leg muscles with dissociated sensory loss in the lumbar and sacral dermatomes. Lower limb reflexes are reduced or absent. Impairment of sphincter function is common.

Other manifestations

Painless ulcers of the hands are frequent. Edema and hyperhidrosis can be due to interruption of central autonomic pathways. Neurogenic arthropathies (Charcot joints) may affect the shoulder, elbow, or wrist [13]. Scoliosis is seen sometimes [14, 15]. Acute painful enlargement of the shoulder is associated with destruction of the head of the humerus.

Physical Examination

A complete physical examination may reveal diminished arm reflexes, which are sometimes present early in the clinical course of syringomyelia. Lower limb spasticity, which may be asymmetrical, appears with other long-tract signs such as paraparesis, hyperreflexia, and extensor plantar responses. Rectal examination includes an evaluation of volitional sphincter control and sensory assessment of sacral dermatomes. Dissociated sensory impairment may be noted. The syrinx may extend into the brain stem, affecting cranial nerves or cerebellar function. Brainstem signs are common in syringomyelia associated with Chiari malformations.

Diagnostic Considerations

Failure to recognize the symptoms of syringomyelia or attributing symptoms (pain, excess sweating, increased spasticity, numbness, or weakness) to other causes can result in morbidity, including neurologic deterioration. Clinicians should have a high index of suspicion for syringomyelia in any patient with SCI presenting with new onset or worsening of any of the aforementioned symptoms. The presence of a peripheral nerve disorder can alter the signs of myelopathy, masking both the sensory loss and distal hyperreflexia. Worsening syringomyelia could be missed in this setting.

Differential Diagnoses may include, 1. Acute Inflammatory Demyelinating, 2. Olyradiculoneuropathy, 3. Amyotrophic Lateral Sclerosis, 4. Atlantoaxial Instability in Down Syndrome, 5. Brainstem Gliomas, 6. Central Pontine Myelinolysis, 7. Chronic Inflammatory Demyelinating Polyradiculoneuropathy, 8. Diabetic Neuropathy, 9.Cervical Spondylosis, 10. Hydrocephalus, 11. Ankylosing Spondylitis, 12. Meningioma, 13. Neural Tube Defects, 14. Neuro-Ophthalmologic Manifestations of Multiple Sclerosis (MS), 15. Pediatric Ependymoma, 16. Pediatric Medulloblastoma, 17. Limb-Girdle Muscular Dystrophy, 18. Spinal Cord Hemorrhage, 19. Spinal Cord Infarction, 20. Spinal Cord Trauma and Related Diseases, 21. Spinal Epidural Abscess, 22. Spinal Metastasis, 23. Spinal Muscular Atrophy and 24. Vascular Surgery for Arteriovenous Malformations

Imaging Studies

Magnetic resonance imaging [16, 17]

Magnetic resonance imaging (MRI) is essential to diagnose syringomyelia. Imaging of the entire rostrocaudal extension of the cyst or cysts is important. Gadolinium-enhanced images are indicated if a tumor is suspected. Gadolinium-enhanced images are helpful in differentiating between scar or disk material associated with a syrinx, especially in postoperative or posttraumatic cases. MRI examination should include sagittal and transverse views in T1 and T2 images. Magnetic resonance angiography can be especially helpful in cases of syringomyelia associated with vascular lesions. Cine phase-contrast MRI is used to analyze CSF flow dynamics near the spinal cord cyst.

Myelography

Myelography is performed in special situations when MRI cannot be used. Widening of the cord and complete subarachnoid block may be observed. Myelogram combined with immediate and delayed high-resolution CT scan also can be performed. Delayed CT scans are obtained 4-24 hours after the initial testing and can demonstrate cyst filling.

Other Tests

In neurophysiological assessment by somatosensory evoked potentials (SSEPs), low-amplitude or delayed responses are present in myelopathy. Neurophysiological assessment by motor evoked response may be more sensitive than SSEPs in the evaluation of spinal cord dysfunction.

Histologic Findings

The syringomyelic cavity, or syrinx, forms most commonly in the lower cervical region, particularly at the base of the posterior horn and extending into the central gray matter and anterior commissure of the cord. Histopathologic findings include (1) cavitation of spinal cord gray matter, (2) syrinx continuous with or adjacent to the central canal, and (3) an inner layer of gliotic tissue. In association with the syrinx, other pathological conditions such as tumors, vascular anomalies, or infective processes also may be evident.

Medical Care

No medical treatment is known for patients with syringomyelia. However, a chronic, stable clinical course is common. Identifying the underlying cause of syrinx formation is very important. Surgical treatment most likely will be necessary. Neuro - rehabilitative care facilitates preservation of remaining neurological functions and prevents complications of quadriparesis such as infection and decubitus ulcers.

Surgical Care

A variety of surgical treatments have been proposed for syringomyelia and are discussed below.

Suboccipital and cervical decompression

This operation includes suboccipital craniectomy; laminectomy of C1, C2, and sometimes C3; and duraplasty [18, 19].

Laminectomy and syringotomy (dorsolateral myelotomy)

After decompression, the syrinx is drained into the subarachnoid space through a longitudinal incision in the dorsal root entry zone (between the lateral and posterior columns), usually at the level of C2-C3. Incision in the dorsal root entry area has the minimum risk of increasing neurological deficit.

Shunts

The following types of shunts may be indicated: 1. Ventriculoperitoneal shunt, 2. Lumboperitoneal shunt - Placed infrequently because of increased risk of herniation through the foramen magnum,3. Syringosubarachnoid dorsal root entry zone shunt and 4. Syringoperitoneal shunt

Terminal ventriculostomy

Neuroendoscopic surgery

This technique is particularly useful in evaluating and treating multiple septate syrinxes. A fibroscope inserted through a small myelotomy allows inspection of the intramedullary cavity. Septa are fenestrated, either mechanically or by laser. Fluid from the cavity is then shunted into the subarachnoid space. Surgical untethering in select cases with posttraumatic tethering associated with syringomyelia [20].

Case report

43 years old female admitted with complaints of head ache, and neck pain of 4 years duration, and abnormal mobility of left shoulder joint &loss of pain sensation in left upper limb of 2 years duration.

History of presenting illness; Head ache of 4yrs duration, lasting for 1-2 hours daily; mostly in morning; not progressive in occipital region, relieved by drugs, increased by coughing, sneezing, working like cooking. Not associated with any diminished visual acuity, no vomiting,

or aura. History of neck pain of 2 years duration, more in left side, Insidious in nature, not progressive. It is of dull aching, continuous, not radiating, and increased while working like cooking because of his she stopped cooking. It was not associated with shock like sensation, the pain is not increased by neck movements. History of abnormal excessive mobility of left shoulder of 2 years, No history of trauma, no history of swelling. While abducting left shoulder she can dislocate and reduces herself voluntarily

There was history of loss of pain and temperature in left upper limb and nape of neck of 2 years duration, she was able to feel clothing, No history of tingling, numbness, no history of weakness. No history of unsteadiness while walking, No history of incoordination in the dark, No history of involuntary movements, No history suggestive of cranial nerve involvement, No history of sweating disturbance, No history of bladder, bowel involvement, No history of seizures.

On examination

Patient was conscious , oriented to time and place, she was afebrile and there was no pallor, jaundice, lymph adenopathy, Height: neck ratio = 11, Upper/ lower segment ratio - 1, height: arm span ratio –normal, No neuro cutaneous markers, No trophic changes in the limbs, No nerve thickening, No digital ulcer.

Vitals; BP Supine - 110/80 mmhg and Standing - 108/80 mmhg, RR- 12/mt, PR -78/ mt

CVS, RS – NAD, CNS –intellectual functions – normal, Cranial nerves –normal.

Motor system

Bulk, power, tone – normal and Superficial reflexes were normal

Deep tendon reflexes	Right	Left
Biceps jerk	+	**Absent**
Triceps jerk	+	+
Supinator jerk	+	**Absent**
Knee jerk	+	+
Ankle jerk	+	+

Plantars – Both Plantar Flexors

Co ordination Test - Normal in all the limbs, No Nystagmus, No Cerebellar Signs, No Involuntary Movements.

Sensory system

Loss of pain and temperature from C3 to T2 in left side

Gait – Normal, No Signs of Meningeal Irritation, Skull – no deformities, Spine – no scoliosis or kyphosis.

No trophic changes, no nerve thickening, no digital ulcer

Both Eyes - Fundus – Normal

Laboratory investigatiuns

CBC – Hb - 12 gms%, PCV - 40, TC - 7000, DC – P - 60, L - 40, ESR - 10 mm 1st hour

RBS - 120 mg%, Urea – 20 mg. S Creatinine - 0.7 mg, Na - 140, K - 4.5, VDRL – Neg, HIV I and II – Neg, Nerve Conduction Study - Normal

MRI (**Figure - 1**) of the head and spine (T2 weighted sagittal view) at time of presentation revealed a cystic lesion involving the cervical spinal cord from the C2 level extending to the C6 level as well as an Arnold-Chiari type 1 malformation. MRI Head (axial view, without contrast) at time of presentation was unremarkable (**Figure – 2**).

Final diagnosis: Syringomyelia involving cervical cord region with Arnold – Chiari malformation type 1.

Discussion

Syringomyelia

It is frequently associated developmental abnormalities

1. In vertebral column (thoracic scoliosis, fusion of vertebrae, or Klippel-Feil anomaly),
2. At the base of the skull (platybasia, basilar invagination),
3. At cerebellum and brain (type I Chiari malformation)

90 percent of cases of syringomyelia have type I Chiari malformation. Syringomyelia is a fluid-filled cavity within the spinal cord. Other nomenclature are Hydromyelia, Syringohydromyelia, Spinal cord cyst

Figure – 1: MRI of the head and spine (T2 weighted sagittal view) at time of presentation revealed a cystic lesion involving the cervical spinal cord from the C2 level extending to the C6 level as well as an Arnold-Chiari type 1 malformation.

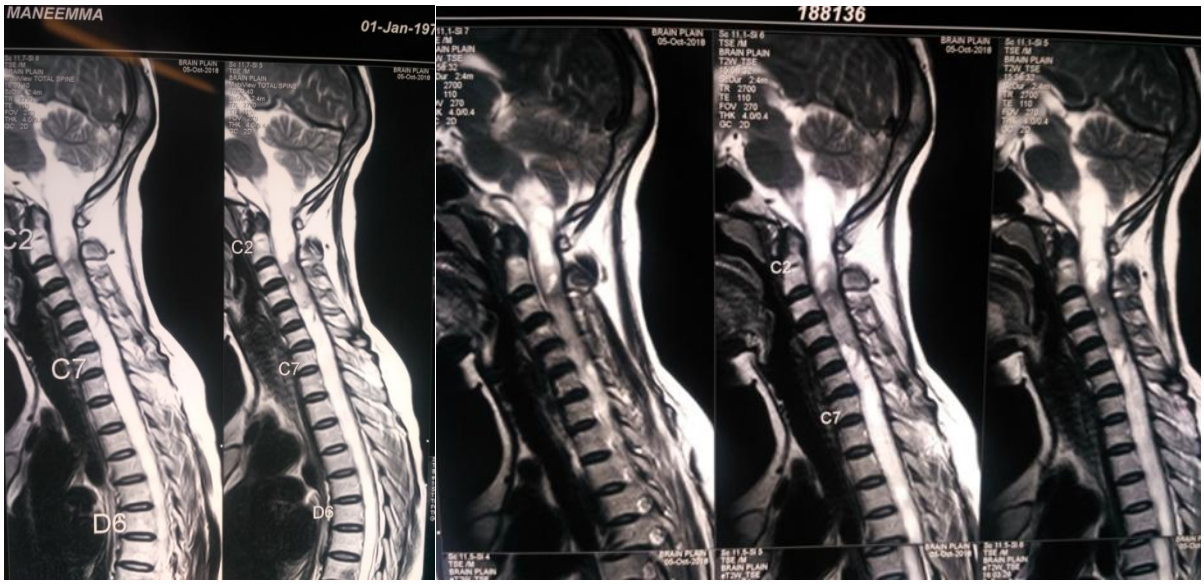
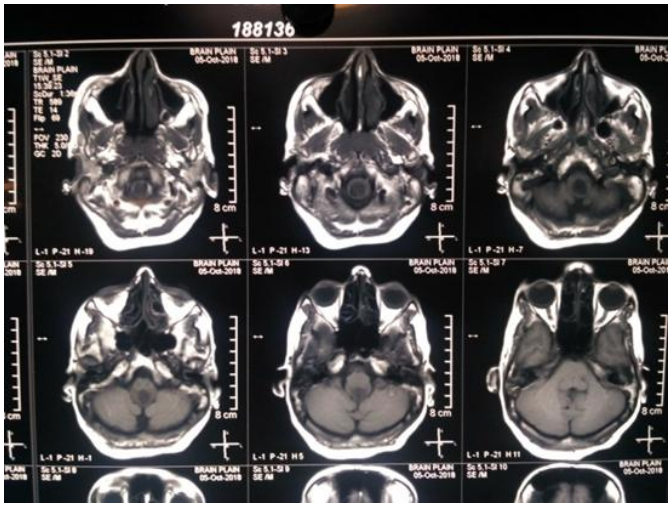


Figure – 2: MRI Head (axial view, without contrast) at time of presentation was unremarkable.



Signs and symptoms

Dissociated sensory loss, the dissociative anethesia takes the Distribution of Short Jacket - Long Sleeve. Because the most common site of the syrnx is C5-T1 Central cord syndrome, Brainstem symptoms and signs, Scoliosis, Chronic pain.

Frequently associated developmental abnormalities include: 1.vertebral column (thoracic scoliosis, fusion of vertebrae, or Klippel-Feil anomaly), 2. base of the skull (platybasia, basilar invagination), 3. cerebellum and brain (type I Chiari malformation)

90 percent of cases of syringomyelia have type I Chiari malformation.

Types

1. Congenital - associated with chiari malformations

2. Acquired – Spinal cord tumors (usually intramedullary, especially hemangioblastoma), Traumatic myelopathy, Spinal arachnoiditis [21] and pachymeningitis, Secondary myelomalacia from cord compression (tumor, spondylosis), infarction, hematomyelia [22]

Idiopathic

Connection with fourth ventricle. Depending on the connection with fourth ventricle, it is described as A. Communicating, B. Non communicating and C. Extra canalicular
Depending on the connection with fourth ventricle Symptoms begins unilaterally Syrinx gradually destroys: 1. decussating S/T tracts, 2. ant. horn cells, 3. lateral C/S tracts,
4. sympathetic tracts and 5. trigeminal, 1X, X, X1 & X11 cranial N nuclei and vestibular system as syrinx extends to the medulla.

Sensory system – There is dissociated sensory loss in either or both arms, or in a shawl like distribution, Dysesthetic pain, a common complaint in syringomyelia, usually involves the neck and shoulders, but may follow a radicular distribution in the arms or trunk. When the cavity enlarges to involve the posterior columns, position and vibration senses in the feet are lost; astereognosis may be noted in the hands.

Motor system - Syrinx extension into the anterior horns of the spinal cord damages motor neurons (lower motor neuron) and causes diffuse muscle atrophy that begins in the hands and progresses proximally to include the forearms and shoulder girdles. Claw hand may develop. There may be Respiratory insufficiency, which usually is related to changes in position, may occur.

Autonomic system - Impaired bowel and bladder functions usually occur as a late manifestation. Sexual dysfunction may develop in long-standing cases. Horner syndrome may appear, reflecting damage to the sympathetic neurons in the intermediolateral cell column.

Extension of the syrinx

a. Syringobulbia [6, 7]

It presents with dysphagia, nystagmus, pharyngeal and palatal weakness, asymmetric weakness and atrophy of the tongue, and loss of pain, temperature in the distribution of the trigeminal nerve.

b. Syringocephalus

Rarely, the syrinx cavity can extend beyond the medulla in the brain stem into the centrum semiovale.

c. Lumbar syringomyelia - it presents with atrophy of the proximal and distal leg muscles with dissociated sensory loss in the lumbar and sacral dermatomes. Lower limb reflexes are reduced or absent. Impairment of sphincter function is common.

Other manifestations - Arm reflex is diminshed or absent. There may be Painless ulcers of the hands. Edema and hyperhidrosis can be due to interruption of central autonomic pathways. There may be Neurogenic arthropathies (Charcot joint). MC shoulder [6] Scoliosis is seen sometimes [12, 13].

Imaging

1.X ray cervical spine - Osseous anomalies of the skull base and skeletal system are observed in 25-50% of pts, Platybasia, basilar invagination (25-50%) , Atlanto-occipital assimilation (1-5%), Klippel-Feil syndrome (5-10%), Incomplete ossification of C1 ring (5%), Proatlantal remnantspina bifida at the C1 level, Retroflexed odontoid process (26%), Scoliosis (42%), Kyphosis, Increased cervical lordosis, Cervical ribs, Fused thoracic ribs.

2.CT scan - CT scanning is reliable in detecting osseous abnormalities, Obliterated cisterna magna, Hydrocephalus, Flattened spinal cord, Tonsillar ectopia, Peg like cerebellar tonsils, Normally positioned fourth ventricle.

3.Spinal MRI will show a dilated cavity with the same intensity of CSF.A complete brain and spinal MRI with and without Gadolinium is needed to determine the primary pathology.

4. MRI – shows the displacement of cerebellar tonsils below the level of the foramen magnum, Pointed and/or peglike tonsils, narrow posterior cranial fossa, elongation of the fourth ventricle, which remains in the normal position. Hindbrain abnormalities, Obstructive hydrocephalus and

associated abnormalities such as syringomyelia and skeletal abnormalities

Tonsillar ectopia

Tonsillar tips that extend less than 3 mm below the landmark are normal. Tonsillar herniation should be primary and not secondary to an intracranial mass lesion to meet the criteria for congenital Chiari I malformation. The most reliable criterion is herniation of at least 1 cerebellar tonsil that is 5 mm or more below the plane of the foramen magnum, Tonsillarectopia of 5 mm is 100% specific and 92% sensitive for Chiari I malformation. Tonsillar herniation of less than 5 mm does not exclude the diagnosis. Herniation of both tonsils that are 3-5 mm below the foramen magnum, accompanied by certain other features, may suggest Chiari I malformation. Cerebellar tonsils usually ascend with age. Some authorities suggest the following criteria for tonsillar ectopia: (1) herniation of 6 mm in those aged 0-10 years, (2) herniation of 5 mm in those aged 10-30 years, (3) herniation of 4 mm in those aged 30-80 years, and (4) herniation of 3 mm in those aged 80-90 years.

Other findings in MRI - Narrowing or obliteration of the retrocerebellar CSF spaces - lower pole of the cerebellar tonsils. The height of supraocciput is reduced, The slope of tentorium is increased. The posterior cranial fossa volume, expressed as a ratio of supratentorial volume (posterior fossa ratio), is significantly smaller; however, mean brain volumes did not differ in patients and control subjects. The cervical subarachnoid space below the level of the C2-3 disks is markedly narrowed in patients with syringomyelia as a result of spinal cord expansion. The posterior subarachnoid space below the tip of the cerebellar tonsils may be completely obliterated.

5. Cine MRI - CSF flow study with phase-contrast cine MRI. Brain pulsations results in caudad and cephalad flow of CSF across foramen magnum during systole and diastole. The reversal in the direction of flow is picked up by alternating light and dark appearance of CSF in front and behind the medulla and upper spinal cord on phase-contrast cine MRI.

CSF flow analysis through foramen magnum with phase-contrast cine MRI helps distinguish symptomatic Chiari I from asymptomatic cerebellar ectopia and helps predict response to surgical decompression.

Treatment

Treatment is based on Etiology

Asymptomatic patients with small syrinx cavity and no obvious etiology are best managed with watchful waiting and serial imaging. Large syrinx: Treat the cause of the syrinx, not the syrinx itself. Analgesics - for headache & neck pain.

Surgery includes decompressive surgery, Suboccipital and cervical decompression. Laminectomy and syringotomy (dorsolateral myelotomy)

Shunts - Ventriculoperitoneal shunt is indicated if ventriculomegaly and increased intracranial pressure are present, Syringosubarachnoid dorsal root entry zone shunt, Syringoperitoneal shunt

Fourth ventriculostomy

Neuroendoscopic surgery

A fibroscope inserted through a small myelotomy allows inspection of the intramedullary cavity. This technique is particularly useful in evaluating and treating multiple septate syrinxes. Septa are fenestrated, either mechanically or by laser. Fluid from the cavity is then shunted into the subarachnoid space.

Operative Results - The most commonly-performed surgery is suboccipital craniectomy (essentially opens up the foramen magnum), with or without C1 laminectomy and dural graft patch. Patients with pain as primary complaint respond best to surgery; weakness less responsive, but overall ~80% of patients report favorable results. Presence of muscle atrophy, ataxia, and duration

of symptoms >2 years all associated with poorer outcome.

Spina bifida

The syrinx may be the result of Tethered cord from the myelomenigocele repair scar,Chiari II malformation, Ventricular shunt malfunction. Location of the syrinx within the spinal cord may help to dictate the treatment

Congenital tethered cord (Spina bifida occulta) - Diagnosed by MRI, treatment is tethered cord release. If syrinx is large, it is often drained at the same surgery

Arachnoiditis - Diagnosis made on MRI, treatment by dissection of the arachnoid scar (often difficult or impossible). Goal is to reestablish normal CSF flow. If the arachnoiditis is so diffuse that it becomes impossible to achieve a good dissection, shunt the syrinx to the pleural or peritoneal cavities.

Trauma - Post-traumatic syrinx is difficult to treat successfully. Possible causes; Arachnoiditis and blockage of flow causing expansion of the cord, or atrophy long term after cord contusion. Treatment is arachnoidal dissection, or syrinx shunt into the pleura or peritoneum.

Spinal cord tumor - Diagnosis made on MRI. High protein content.

Treatment is by tumor resection. It is rare to have to shunt the syrinx in these situations.

Idiopathic (No identifiable cause) - In a large percentage of patients, the syrinx has no identifiable cause.it is difficult to treat. If large, syrinx shunting is done. Rarely, posterior fossa decompression (Chiari zero). It is so far impossible to predict which patient with idiopathic syringomyelia would respond to posterior fossa decompression

Prognosis and outcome (Syringomyelia resolution)

Chiari decompression - Excellent outcome. Spina bifida - Excellent outcome when shunt is functional, Arachnoiditis–if it is focal – good prognosis, if it is diffuse – poor prognosis, need to shunt the syrinx, Trauma - Poor outcome for syringomyelia and pain

Tumor - Excellent outcome for syringomyelia, overall Prognosis depends on tumor grade.

Arnold–Chiari malformations

Arnold–Chiari malformations, types I-IV, refer to a spectrum of congenital hindbrain abnormalities affecting the structural relationships between the cerebellum, brainstem, the upper cervical cord, and the bony cranial base.

Type I - A congenital malformation. Most common herniation of cerebellar tonsils Syndrome of occipitoatlantoaxial hypermobility. It is an acquired Chiari I Malformation in patients with hereditary disorders of connective tissue. Patients exhibit extreme joint hypermobility and connective tissue weakness as a result of Ehlers-Danlos syndrome or Marfan Syndrome [22]. And they are susceptible to instabilities of the craniocervical junction and thus acquiring a Chiari Malformation. This type is difficult to diagnose and treat.

Type I - True incidence –not known, male to female ratio - 2:3. It is common in adult and paediatric age group. Incidence syrinx- 25—70%. syringohydromyelia is secondary to pathologic CSF dynamics

SYMPTOMS – symptoms are due to disruption of CSF flow through foramen magnum. Chiari I Malformation symptom are head ache. Headache and neck pain in Chiari I are often exacerbated by cough and Valsalva manoeuvre. In syringomyelia and central cord symptoms such as hand weakness and dissociated sensory loss is present

Symptoms are due to 1. Compression of medulla and upper spinal cord, which include myelopathy, lower cranial nerve palsies, and nuclear dysfunction. 2. Compression of cerebellum which include ataxia, dysmetria, nystagmus, dysequilibrium.

William's theory
herniated tonsil at foramen magnum acts like a valve. The Pressure differrence increases. The increase in subarachnoid fluid pressure from increased venous pressure during coughing or Valsalvamaneuvers is localized to the intracranial compartment. It increases the cisterna magna pressure simultaneously with a decrease in spinal subarachnoid pressure. This craniospinal pressure gradient draws CSF caudally into the syrinx.

New concept
In chiari malformation, the pressure in veins and capillary around central canal is very high. Coughing , sneezing , even heart beat put more stress on blood vessels, and causes leakage of plasma – form syrinx .

Type II - Usually accompanied by a lumbar myelomeningocele leading to partial or complete paralysis below the spinal defect - a larger cerebellar vermian displacement. Low lying torcularherophili, tectalbeaking, and hydrocephalus with consequent clival hypoplasia

Type III - Causes severe neurological defects. It is associated with an occipital encephalocele.

Type IV - Characterized by a lack of cerebellar development.

Causes - May be Genetic with chromosome 9&15 involvement or Vitamin deficiencies

Conclusion

Syringomyelia is the development of luid-filled cavity or syrinx within the spinal cord. It is associated with craniovertebral junction abnormalities. Syrinx interrupts the decussating spinothalamic fibers that mediate pain and temperature sensibility, resulting in loss of these sensations, while light touch, vibration, and position senses are preserved (dissociated sensory loss).Natural history of syringomyelia still is not well understood.MRI examination should include sagittal and transverse views in T1 and T2 of the entire rostrocaudal extension of the cyst or cysts is important.

No medical treatment is known for patients with syringomyelia. The surgical treatment will be necessary. A variety of surgical treatments for syringomyelia which are available are described. We herewith report a rare case of syringomyelia and associated Chiari I malformation presenting with dissociated sensory impairment in neck region with headache and neck pain Treatment in these cases is surgical decompression. Recovery with significant decrease in sensory loss and relief in headache and neck pain.

References

1. Nakanishi K, Uchiyama T, Nakano N, et al. Spinal syringomyelia following subarachnoid hemorrhage. J Clin Neurosci., 2012 Apr.; 19(4): 594-7.
2. Bethesda MD. Syringomyelia Fact Sheet. National Institute of Neurological Disorders and Stroke; 2011. NIH Publication No., 10–3780.
3. Srivatanakul K, Songsaeng D, Ozanne A, Toulgoat F, Lasjaunia P. Spinal arteriovenous malformation associated with syringomyelia. J Neurosurg Spine, 2009; 10(5): 436–42.
4. Kim J, Kim CH, Jahng TA, Chung CK. Clinical course of incidental syringomyelia without predisposing pathologies. J ClinNeurosci., 2012; 19(5): 665-8.
5. Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. J Neurol Neurosurg Psychiatry, 1965; 28: 247-59.
6. Williams B. Progress in syringomyelia. Neurol Res., 1986; 8(3): 130-45.
7. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. J Neurosurg., 1994; 80(1): 3-15.
8. Kiran NA, Kasliwal MK, Suri A, Mahapatra AK. Giant posterior fossa arachnoid cyst associated with

syringomyelia. Clin Neurol Neurosurg., 2010; 112(5): 454-5.
9. Van Gijn J, Giesbergen PCLM, Hoogengraad TU. Attacks of pain in the leg from classic syringomyelia. J Neurol Neurosurg Psychiatry, 1997; 63(3): 410.
10. Sixt C, Riether F, Will BE, Tatagiba MS, Roser F. Evaluation of quality of life parameters in patients who have syringomyelia. J Clin Neurosci., 2009; 16(12): 1599-603.
11. Viswanatha B. Syringomyelia with syringobulbia presenting as vocal fold paralysis. Ear Nose Throat J., 2009 Jul; 88(7): E20.
12. Tubbs RS, Bailey M, Barrow WC, Loukas M, Shoja MM, Oakes WJ. Morphometric analysis of the craniocervical juncture in children with Chiari I malformation and concomitant syringobulbia. Childs Nerv Syst., 2009; 25(6): 689-92.
13. Nacir B, Arslan Cebeci S, Cetinkaya E, Karagoz A, Erdem HR. Neuropathic arthropathy progressing with multiple joint involvement in the upper extremity due to syringomyelia and type I Arnold-Chiari malformation. Rheumatol Int., 2010; 30(7): 979-83.
14. Ono A, Suetsuna F, Ueyama K, Yokoyama T, Aburakawa S, Numasawa T. Surgical outcomes in adult patients with syringomyelia associated with Chiari malformation type I: the relationship between scoliosis and neurological findings. J Neurosurg Spine, 2007; 6(3): 216-21.
15. Cardoso M, Keating RF. Neurosurgical management of spinal dysraphism and neurogenic scoliosis. Spine (Phila Pa 1976), 2009; 34(17): 1775-82.
16. Aubin ML, Vignaud J, Jardin C, Bar D. Computed tomography in 75 clinical cases of syringomyelia. Am J Neuroradiol., 1981; 2(3): 199–204.
17. Brodbelt AR, Stoodley MA. Post-traumatic syringomyelia: a review. J Clin Neurosci., 2003; 10(4): 401–8.
18. Lucchetta M, Cagnin A, Calderone M, Manara R, Rotilio A, Briani C. Syringomyelia associated with Chiari I malformation. Neurol Sci., 2009; 30(6): 525-6.
19. Prat R, Galeano I. Pain improvement in patients with syringomyelia and Chiari I malformation treated with suboccipital decompression and tonsillar coagulation. J Clin Neurosci., 2009; 16(4): 531-4.
20. Falci SP, Indeck C, Lammertse DP. Posttraumatic spinal cord tethering and syringomyelia: surgical treatment and long-term outcome. J Neurosurg Spine, 2009 Oct.; 11(4): 445-60.
21. Hamlat A, Adn M, Yahia MB, Morandi X, Brassier G, Guegan Y. Gowers intrasyringal hemorrhage case report and review of the literature. J Neurosurg Spine, 2005; 3(6): 477–81.
22. Banerji NK, Millar JHD. Chiari malformation presenting in adult life. Its relationship with syringomyelia. Brain, 1974; 97(1): 157–68.