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A REVIEW ON SYRINGOMYELIA

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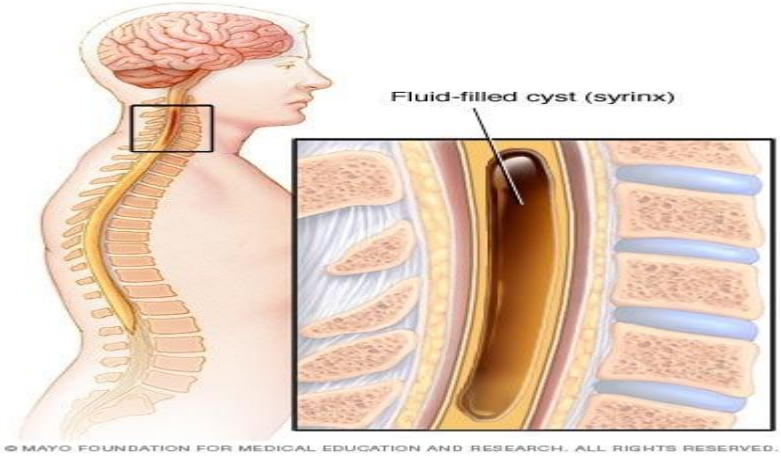
ABSTRACT

Syringomyelia classically presents as a bilateral sensory loss of the dissociated type, including the loss of pain and temperature with the preservation of fine touch, vibratory sensation, and proprioception in the shoulder, arm, and hand. Eventually, weakness of the legs, muscle wasting, and ataxia can also be seen due to the involvement of the corticospinal tracts and the posterior columns of the spinal cord. We present the case of a 64-year-old patient with an atypical presentation of post- traumatic syringomyelia. This atypical presentation included a unilateral sensory loss of fine touch, pain, and temperature in the shoulder, arm, and hand which was of the non-dissociated type with no weakness, muscular atrophy, loss of vibratory sensation, or proprioception. Syringomyelia is a chronic, progressive disease of the spinal cord. Syringomyelia is an etiologically diverse affliction caused by disturbance of normal cerebro spinal fluid flow dynamics. Lesions are characterized by the formation of tubular cavities in the gray matter of the spinal cord and gliosis; however, the aetiology is unknown and treatment methods differ. Many existing studies have focused on the relationship between other diseases and syringomyelia. There is a lack of comprehensive and objective reports on the research status of syringomyelia. Therefore, this study aimed to conduct a bibliometric analysis to quantify studies on Syringomyelia and trending issues in the last 20 years. In the pre-MR era syringomyelia often presented late, as a crippling neurological disorder.

KEYWORDS: Syringomyelia, Treatment, Cerebrospinal Fluid.

INTRODUCTION



Syringomyelia is a neurological disorder in which a fluid filled cyst(syrinx) forms within the spinal cord. The syrinx can get big enough to damage the spinal cord and compress and injure the nerve fibres that carry information to and from the brain to the body. Syringomyelia predominately presents with sensory symptoms such as pain and temperature insensitivity. It may causes in paraplegia and quadriplegia. The syrinx is a result of disrupted CSF drainage from the central canal, the syrinx may occur in the brainstem, and is then referred to as syringobulbia. The syrinx initially compress and permanently damages crossing fibres of the spinothalamic tract. Further expansion may affect other tracts of the spinal cord. Symptoms include dissociated sensory loss, Symptoms include dissociated sensory loss, presenting as cape like distribution of decreased sensitivity to pain and temperature and flaccid atrophic paralysis in the upper extremities. Syringomyelia typically seen in conjuction with type 1 chairi malformation (CMS)Other known etiologies include spinal cord tumours, trauma and post traumatic or infections adhesive arachnoiditis.

Although syringomyelia may present with sensory symptoms such as pain and temperature insensitivity, it is more commonly found incidentally. The increased utilization of magnetic resonance imaging (MRI) for the evaluation of neck or back pain has led to increased detection of syringomyelia. Syringomyelia accounts for up to 5 percent of paraplegia. This activity addresses the presentation, evaluation, and management of syringomyelia and examines the role of an interprofessional team approach to the care of affected Over time, the cyst can enlarge, damaging your spinal cord and causing pain, weakness and stiffness, among other symptoms.

Syringomyelia has several possible causes, though the majority of cases are associated with a condition in which brain tissue protrudes into your spinal canal (Chiari malformation). If syringomyelia doesn't cause problems, monitoring the condition might be all that's necessary. But if you're bothered by symptoms, you might need surgery. In syringomyelia, the watery liquid known as cerebrospinal fluid (CSF)which surrounds and protects the brain and spinal cord builds up within the tissue of the spinal cord, expands the central canal, and then forms a syrinx. Generally, a syrinx develops when the normal flow of CSF around the spinal cord or lower brain stem is disturbed. When syrinxes affect the brain stem, the condition is called syringobulbia. symptoms of damage to the spinal cord vary among individuals depending on where the syrinx froms, how long it extends. Symptoms develops slowly over time, worsen over many years, and may occur on one or both the sides of the body. You may not have symptoms, or even be aware that you have it. But a severe case symptoms depend on the location and size of the cyst. over time, if it grows wider and longer, it can damage the nerves in the centre of the spinal cord.

HISTORY

Syringomyelia is a composite from two Greek words Syrinx and Myeols. The disease was first coined by Charles-prosper Oliver d' Angers in 1827. but the condition was first recognized by Stephanus in 1545. Schultze first described its clinical picture; its relation to chiari malformation was first outlined by John Cleland. Abbe performed the first reported surgical drainage of a syrinx in 1891, although claims were made that Horsley aspirated a syrinx in 1890. The "hydrodynamic" theory of syringomyelia was proposed by Gardner in 1965 and modified by Williams in 1978 with the "craniospinal pressure dissociation".

TYPES

There are broadly three types of syringomyelia. The most common type is associated with congenital (from birth) brain abnormalities, while the second type develops as a complication following spinal cord, infection or tumors. The third type is caused by unknown factors and is called as idiopathic syringomyelia.

CLASSIFICATION

1) Congenital
2) Acquired
3) Idiopathic

1) CONGENTIAL SYRINGOMYELIA

It is also known as communicating Syringomyelia. It is often caused by Chiari malformation and resulting syrinx usually in the spines cervical (neck) region. Symptoms typically begin between ages 25 and 40 People with congenital syringomyelia may also have hydrocephalus, a buildup of excess CSF in the brain along with larger than normal connected cavities called ventricles. Straining or coughing can increase the pressure within your head and brain, causing you to develop a headache or even lose consciousness. Some people may have a disorder called arachnoiditis an inflammation of the arachnoid, one of the three membranes that surrounds the spinal cord.

2) ACQUIRED SYRINGOMYELIA

• Spinal cord injury

• Meningitis (an inflammation of brain and spinal cord membranes usually caused by an infection)

• Arachnoiditis

• Tethered cord syndrome (a condition present at birth that causes the spinal cord to abnormally attach to the tissues in the lower spine, limiting its movement)

• Spinal cord tumor.

3) IDIOPATHIC

Idiopathic syringomyelia is a rare, non-syndromic central nervous system malformation characterized by a longitudinally oriented fluid-filled cavity inside the spinal cord parenchyma or the central canal, without any readily identifiable cause. It is usually associated with pain, sensory and/or musculoskeletal disturbances, but it can also be an incidental and asymptomatic finding. Alteration in the cerebrospinal fluid flow dynamics in the region of craniovertebral junction related to chronic inflammation is the most frequently speculated cause of such syringomyelia. As no definite cause is identified, the treatment recommended is usually shunt surgery that involves drainage of the syrinx cavity. There have been reports where opening of the foramen magnum and lysing the arachnoidal adhesions have been recommended. However, no universally accepted treatment protocol is yet available.

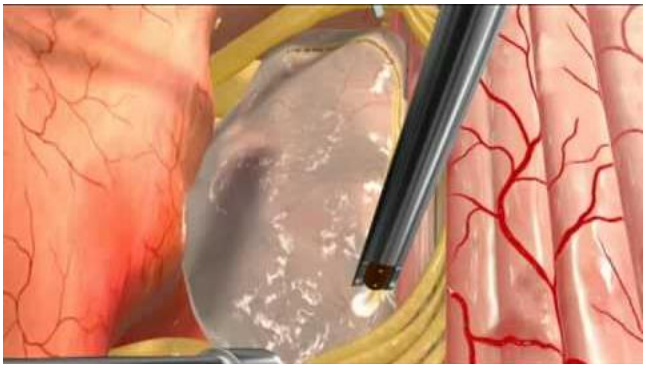
MECHANISM OF ACTION

We know that in syringomyelia, syrinx means cyst or cavity myelia means spinal cord that means cystic enlargement of spinal cord. cerebro Spinal fluid is a clear colourless body fluid

found within the tissues that surrounds around the brain and spinal cord of all vertebrates. CSF is produced by a structure called choroid plexus in the lateral, third and fourth ventricular of the brain. From the fourth ventricular it passes to the spinal cord. The cavity starts middle of intracanal expands out and damages the spinothalamic tract. Cerebrospinal fluid circulates through a system of cavities found within the brain and spinal cord; ventricles, subarachnoid space of the brain and spinal cord and the central canal of the spinal cord. The secretion of CSF equals its removal, so there is around 150-270 millilitres of cerebrospinal fluid within the CNS at all times.

The main functions of CSF are to cushion the brain and spinal cord when they're struck with mechanical force, to provide basic immunological protection to the CNS, to remove metabolic waste, as well as to transport neuromodulators and neurotransmitters. CSF is also very useful for clinical diagnosis, and its samples are usually obtained from the subarachnoid space (SAS) by lumbar puncture. Syrinxes usually result from lesions that partially obstruct flow of cerebrospinal fluid (CSF). At least half of syrinxes occur in patients with congenital abnormalities of the cranio cervical junction (eg herniation of cerebellar tissue into the spinal canal, called Chiari malformation), brain (eg encephalocele), or spinal cord (eg myelomeningocele). For unknown reasons, these congenital abnormalities often expand during the teen or young adult years.

A syrinx can also develop in patients who have a spinal cord tumor, scarring due to previous spinal trauma, or no known predisposing factors. About 30% of people with a spinal cord tumor eventually develop a syrinx. The Spinal cord itself is composed of gray and white matter, Gray matter is found within the middle portion of spinal cord and it's shape is like butterfly in this where the cell bodies and neurons are present. A syrinx can also develop in patients who have a spinal cord tumor, scarring due to previous spinal trauma, or no known predisposing factors. About 30% of people with a spinal cord tumor eventually develop a syrinx. The Spinal cord itself is composed of gray and white matter, Gray matter is found within the middle portion of spinal cord and it's shape is like butterfly in this where the cell bodies and neurons are present. In the center of gray matter there is a small cavity called the central canal which is filled with cerebro spinal cord. In white matter consists of axons of neurons that carry Information from the brain.



The main cause of syringomyelia is genetic condition called chiari malformation type 1. In chiari malformation the displacement of cerebellum and blocking the opening of CSF exits into subarachinoid space then it causes fluid backs up into the spinal cord then it leads to syringomyelia. There are several types of Chiari malformations, but type I is the most common. In type I, the cerebellum bulges through the normal opening at the base of the skull. This type is most often congenital. It is also called primary Chiari malformation type I. But it is often not found until a person is a teen or young adult.

PATHOPHYSIOLOGY

Over the years, numerous theories have attempted to explain the pathogenesis of syringomyelia. We still lack a theory capable of explaining all the possible scenarios that may cause syringomyelia. Although the intramedullary pulse pressure theory proposed by Greitz seems to fulfil this goal, it is yet to be universally accepted; some researchers continue to support other theories that have already been rejected by the scientific community. Traditional and recent theories agree that syringomyelia results from abnormal CSF circulation within the spinal subarachnoid space. Early theories postulated that abnormal CSF circulation was caused by spinal trauma secondary to an ischaemic lesion, leading to spinal cystic degeneration and the formation of a large cavity. According to other theories, atrophy of the spinal cord parenchyma causes the central canal to widen and force the remaining parenchyma outward, compressing CSF circulation through the subarachnoid space. The CSF would then be unable to travel down the spinal cord and would therefore enter the parenchyma, accumulating and forming a fluid-filled cavity. These theories prevailed for a long time but are no longer supported as they are physically implausible. If CSF flow stopped, liquid would accumulate above the obstruction, increasing the pressure in that area and leading to CSF accumulation outside the spinal cord, which would compress the parenchyma. Other outdated theories have suggested a congenital origin associated with

defects in differentiation and neural tube closure; according to the theories postulated by Gardner and Williams, syringomyelia is caused by alterations in CSF circulation in the fourth ventricle or the foramen magnum; Heiss and Oldfield propose a similar theory in the context of Chiari malformation, according to which the movement of the cerebellar tonsils has a piston-like effect.

ETIOLOGY

The etiology of syringomyelia includes conditions that alter the physiologic CSF circulation dynamics. In most cases, it is secondary to spinal subarachnoid space obstruction. Etiology includes.

• Idiopathic Syringomyelia

Syrinx in the absence of an identifiable cause

• Secondary Syringomyelia.

• Syringomyelia with obstruction at the foramen magnum (developmental).

• Chiari 1 Malformation (CM1): Most common association.

• Basilar invagination Syringomyelia with other diseases of the spinal cord (acquired).

• Post-inflammatory Post-infectious: Granulomatous (tuberculosis, fungal), post-operative meningitis Chemical/ Sterile inflammation: Post-subarachnoid hemorrhage (SAH), post myelography(metrizamide).

• Post-traumatic.

• Spinal Cord Tumors: Intra-medullary spinal tumors esp. Hemangioblastoma.

• Secondary myelomalacia: Cord compression (herniated disc, spondylosis, tumors), infarction, hematomyelia.

EPIDERMIOLOGY

Estimated prevalence of the disease is about 8.4 cases per 100,000 people and occurs more frequently in men than in women. The disease usually appears in the third or fourth decade of life, with a mean age of on set of 30 years. Syringomyelia may develop in childhood or late adult hood. The nationwide epidemiological survey of syringomyelia was carried out in Japan by sending inquiries to neurologists, child neurologists, neurosurgeons and Orthopedic surgeons for the period of 1991 and 1992. A total of 1,243 cases of syringomyelia were ascertained. Among them, 622 were men and 619 women, and the average age of onset was 28 years old. The classification by Barnett et al was used, presenting syringomyelia with Chiari malformation in 684 cases (51.2%), dysraphism in 47 (3.7%), post traumatic

syringomyelia in 139 (11%), post-spinal arachnoiditis in 76 (6%), spinal cord tumor in 132 (10.5%) and others in 204. Its predominant clinical course was slowly progressive, but 202 cases (17.9%) showed rather stable course including spontaneous resolution in 29 cases. The main initial symptoms were numbness in 522 cases (42%), motor disturbance in 504 (40.5%), and pain in 296 (23.8%).

SINGS AND SYMPTOMS

The symptoms vary from one person to the next, depending on the severity and location of the cyst, and the underlying cause. Symptoms can develop slowly and this may delay the diagnosis.

Symptoms include: Pain (potentially chronic)

• Progressive weakness in arms and legs
• Stiffness in the back, shoulders, neck, arms, or legs
• Headaches
• Loss of sensitivity to pain or hot and cold, especially in the hands
• Numbness or tingling
• Loss of balance
• Loss of bowel and bladder control
• Curvature of the spine (scoliosis); may be the only symptom in children Muscle weakness and wasting (atrophy)
• Loss of reflexes
• Loss of sensitivity to temperature
• reduced skin sensations.
• Muscle atrophy (wasting), usually beginning in the hands and spreading to include the arms and shoulder
• sexual dysfunction.
• Increased sweating
• Blood pressure changes

CAUSES

It's unclear how and why syringomyelia happens. When it develops, cerebrospinal fluid that surrounds, cushions and protects your brain and spinal cord collects within the spinal cord itself, forming a fluid-filled cyst (syrinx). Several conditions and diseases can lead to syringomyelia, including.

• Chiari malformation, a condition in which brain tissue protrudes into your spinal canal.
• Meningitis, an inflammation of the membranes surrounding your brain and spinal cord.
• Spinal cord tumor, which can interfere with the normal circulation of cerebrospinal fluid.
• Conditions present at birth, such as a tethered spinal cord, a condition caused when tissue attached to your spinal cord limits its movement.
• Spinal cord injury, which can cause symptoms months or years later.

PREVENTION

• Avoid doing anything that worsens your symptoms.
• People with syringomyelia, heavy lifting and straining can trigger symptoms.
• Avoid flexing your neck and jumping.
• Chiari malformation you may experience headache with straining.

PROGNOSIS

SM is a slowly progressing chronic disease, and its prognosis depends on etiology, location, size of the syrinx and the severity of neurological dysfunction. Individuals aged greater than 30 years and who have cord injury are expected to develop PTS within 5 years of the injury. Delayed treatment or undiagnosed SM may lead to irreversible injury in the spinal cord. In patients with mild neurological problems, response to treatment is reported to be satisfactory.100 In 90% of reported cases, syrinx size has been typically reduced after PFD within 6 months.

COMPLICATION

Besides general complications in neurosurgery such as anesthetic and general complications, infections, and post operative hematomas, the most prominent problems in syringomyelia surgery are scar formation around the spinal cord and any implanted shunts, wound leakage of CSF, and shunt obstruction. For direct drainage of a syrinx, a micromyelotomy is required. In paraplegic or tetraplegic patients, this is performed below the functional level so no additional neurological deficit will be made.

CLINICAL PRESENTATION

Pain in the neck, shoulders and hand are the more prominent and constant presenting clinical features. Wasting and weakness of hands and spasticity and weakness in the legs can be present in more longstanding cases. Classically described sensory loss that includes affection of posterior column sensations and affection of the temperature and pain sensations in the

shoulders and hands are more characteristic presenting symptoms. Despite the presence of syringomyelia and reduction in the neural girth, the symptoms are remarkably less and few.

RISK FACTORS

Some features may be regarded as risk factors for syringomyelia in certain groups. Among iatrogenic risk factors, for example, greater quantities of blood at the surgical site or multiple traumatic punctures are associated with increased risk of fibrosis, which may block CSF circulation. The risk factors for post-traumatic syringomyelia are the most frequently studied. Presence of complete spinal cord injury is probably the most significant risk factor. el Masry and Biyani and Curati et al. suggest that complete spinal cord lesions (grade A on the American Spinal Injury Association Impairment Scale) double the risk of clinical syringomyelia. Other known risk factors for this type of syringomyelia are spinal canal stenosis >25% and post-traumatic kyphosis >15°. Presence of these risk factors does not imply a need for surgery to prevent the development of syringomyelia.

DIAGNOSIS

MRI. An MRI of your spine and spinal cord is the most reliable tool for diagnosing syringomyelia. An MRI uses radio waves and a strong magnetic field to produce detailed images of your spine and spinal cord. IF a syrinx has developed within your spinal cord, your doctor will able to view it on the MRI. In some cases a specialist will inject a dye into a blood vessel in your groin, which travels through blood vessels to your spine and reveals tumors or other abnormalities.

CT scan. A CT scan uses a series of x rays to create a detailed view of your spine and spinal cord. It can reveal tumors or other spine conditions.

TREATMENT

- Surgery
- Treating chiari malformation
- Draining the syrinx
- Removing the obstruction Surgery: The goal of surgery for syringomyelia is to eliminate the syrinx and prevent further spinal cord injury. Surgery is usually recommended for cases of symptomatic or progressive syringomyelia. There are two general forms of treatment: restoration of normal CSF flow around the spinal cord, and directly draining the syrinx. The type of treatment depends on what is causing your symptoms.

Treating the Chiari malformation: The main goal of Chiari surgery is to provide more space at the base of your skull and upper neck. This reduces pressure on the brain and spinal cord, restoring the normal flow of CSF. Surgery can allow the syrinx to drain, sometimes becoming smaller or even disappearing entirely. Symptoms may improve even if the syrinx remains the same size or is reduced only slightly. You should get treated sooner rather than later because delaying treatment can cause irreversible spinal cord damage. Syringomyelia can reoccur after surgery, making additional operations necessary.

Draining the syrinx: If there is no associated Chiari malformation or tumor, it may be necessary to drain the syrinx. This is usually done in cases where the syrinx is growing, or the cause of the CSF obstruction cannot be identified. A surgeon will insert a drain called a stent or shunt to allow the fluid to flow to another part of your body where it can be absorbed. Draining the syrinx can halt the progression of symptoms and relieve headache.

Removing the obstruction: Surgically removing obstructions such as scar tissue, bone from the spinal canal, or tumors can help restore the normal flow of CSF. If a tumor is causing your syringomyelia, removing it almost always eliminates the syrinx. Occasionally, radiation may be used to shrink the tumor.

CONCLUSION

Syringomyelia is a rare in our population. It affects young patients, typically in the cervical thoracic region. Early diagnosis and surgical treatment syringomyelia is essential to prevent further loss of neurological function, Restoration of normal CSF flow from the fourth ventricle to sub arachinoid space and relief of direct brainstem compression are the good of surgery, surgeons should not to operate on a sympatomatic patients with small syrinx. A static or transiently increasing syrinx after untethering usually does not imply failed surgery. Clinical presentations of patients with Chiari malformation with or without syringomyelia can be extremely diverse and heterogenous. The understanding of the disease and its pathogenesis is still evolving. All patients with a syrinx regardless of the size, location, or other associated symptoms are offered surgical intervention. Foramen magnum decompression with small sub occipital craniectomy, removal of posterior arch of C1 and duro plasty using G patch graft, with water tight dural closure has given good results.

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