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Case Report

Primary Spinal Syringomyelia with Hemianhidrosis: A Rare Case Report

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ABSTRACT

Background: The incidence of syringomyelia is very rare, at 8.4 per 100,000 people, or about 2%. It is characterized by a cyst filled with cerebrospinal fluid (CSF) within the spinal cord tissue or central canal. Symptoms may include sensory and motor disturbances. **Objective:** We report a case of a 35-year-old woman who presented with pain, weakness, and stiffness in her back, shoulders, arms, and legs. **Methods:** She experienced weakness on her right side for the past five years and had no sweating on that side for two years. Motor examination showed flaccid hemiparesis and muscle atrophy in her right upper limb. Reflex testing revealed hyperreflexia in her right hand and leg. Sensory testing (touch, temperature, pain, vibration) showed dissociation of sensation on the medial-lateral side of her right arm and leg. **Results:** After 1.5 years of treatment, including routine Short Wave Diathermy (SWD), Transcutaneous Electrical Nerve Stimulation (TENS), physical therapy, and medication, her condition improved, and her quality of life was enhanced. **Conclusion:** This case highlights that a multidisciplinary approach, including SWD, TENS, physical therapy, and medication, can effectively reduce symptoms and improve quality of life in syringomyelia patients, even with extensive spinal involvement (C1-T11).

Keywords: cerebrospinal fluid, hemiparesis, muscle atrophy, spinal cord, syringomyelia

1. Introduction

Syringomyelia is a cyst filled with cerebrospinal fluid (CSF) in the parenchyma of the spinal cord or central canal. This disease is classified as a rare neurological condition. The incidence of syringomyelia is very rare at 8.4/100,000, or only 2% [1]. Other studies have shown that the estimated prevalence of syringoma ranges from 1.9 to 5.9 per 100,000. Incidence in men is the same as in women and generally occurs at the age of 25-40 years. The condition can be progressive, causing disability for the sufferer [1,2]. Syringomyelia is also defined as the formation of a fluid-filled cavity called the syrinx, which can enlarge within the spinal cord. Patient complaints vary depending on the location (central, paracentral, or eccentric cavities), size, and length of the spinal cord. Symptoms can include pain, weakness, and stiffness in the back, shoulders, arms, and legs. Other symptoms include headache and loss of sensation of heat or cold, especially in the hands [3] [4].

Based on the cause, Syringomyelia is grouped into primary syringomyelia, which is due to genetic or idiopathic disorders, and secondary Syringomyelia (associated with various conditions). Most cases of syringomyelia are associated with abnormalities of the craniocervical junction, a history of surgery with profuse bleeding, and spinal trauma. Other conditions include spinal dysraphism, intramedullary and extramedullary tumors, and arachnoiditis. Very few cases occur without a known associated pathology [2].

The diagnosis of syringomyelia is determined through anamnesis, physical examination, and supporting examination, namely, Magnetic Resonance Imaging (MRI) of the spinal cord segment. Syringomyelia management can be conservative or operative, depending on the location, size of the lesion, and the development of symptoms experienced by the patient.

2. Case Report

A 35-year-old woman, working as a laborer, from Baypas Padang, came with the chief complaint of weakness in the right side of her limb. The complaint started with stiffness and pain in the head and neck radiating to the left hand. The complaint has been going on for about 5 years, but has been ignored. The longer his left arm could not feel the heat or cold, and it weakened. Sometimes the patient complains of stinging when exposed to the sun, the left shoulder feels sore, and the muscles of both arms feel thinner. In addition, the patient feels that the left side is sweaty when she is doing activities, but the right side of her body does not sweat at all. There was no history of previous trauma, infection, or surgery. There is no family history of malignancy or similar complaints.

In general physical examination, vital signs were found: blood pressure 110/70 mmHg, pulse 77x/, temperature 36.5°C. On neurological examination, the muscles of the arms and fingers atrophy, but tone was still good. Stiffness and tenderness at cervical trigger points. Motor examination revealed hemiparesis of the flaccid type, on the superior and inferior extremities on the left side. Reflex examination revealed a positive Hoffmann-Trommer. Examination of physiologic reflexes showed hyperreflexia of the left hand and leg. Sensibility examination (touch, temperature, pain, and vibration) showed dissociation of sensibility in the right arm and right leg on the medial-lateral side and muscle atrophy on the right arm. Complete blood counts, electrolytes, kidney function, and lipid profiles were performed, and the results were within normal limits. On investigation, cervical MRI (Magnetic resonance imaging (MRI)) of the brain and cervical spine were planned for the patient, and the results are below.

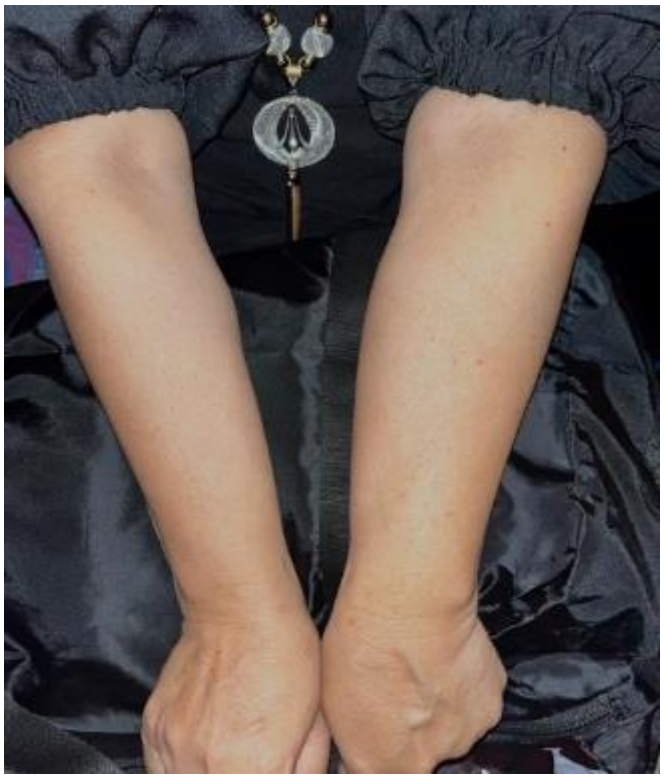

Figure 1. The right hand looks smaller than the left side

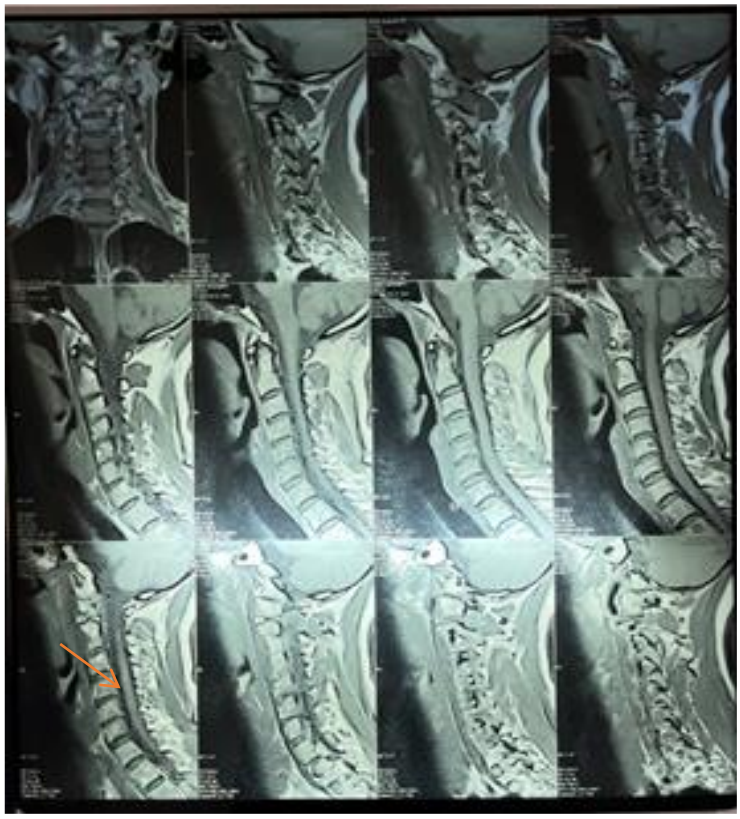

Figure 2. Syringomyelia in the Cervical Region

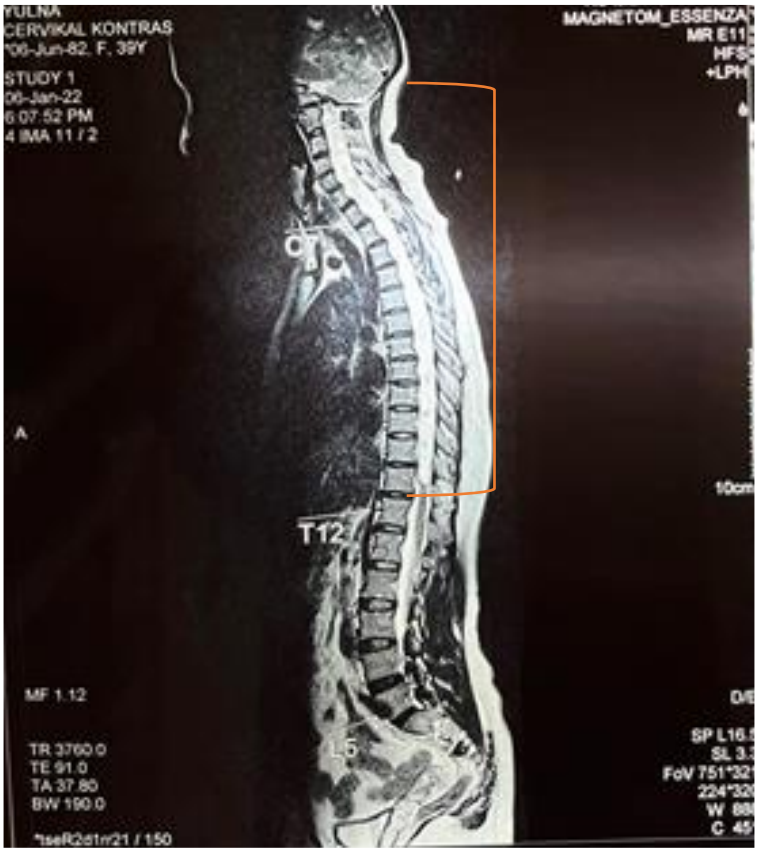

Figure 3. MRI findings of syringomyelia from C1-T11.

After doing physical and supporting examination above, the patient was referred to the orthopedic department, but the patient refused. Therefore, he patient was consulted for medical rehabilitation. The patient received Infrared, Short Wave Diathermy (SWD) and Transcutaneous Electrical Nerve Stimulation (TENS) therapy and Physical Therapy for 1.5 years.

3. Discussion

The majority of patients with syringomyelia are between 20 and 50 years of age and present with decreasing sensory and motor complaints [2]. In adults, the primary etiology of syringomyelia is Chiari I malformation; other causes include trauma, neoplasm, and inflammatory conditions. Current theories regarding the pathogenesis of syringomyelia primarily emphasize hydrodynamic mechanisms, which can be categorized into those that involve flow from the fourth ventricle into the central canal, mechanisms whereby fluid flows across the spinal cord parenchyma from the subarachnoid space, mechanisms that limit outflow, and pressure effects on the spinal cord leading to dissection of the cord tissue by an existing syrinx. In this context, the patient had no history of tumor, infection, or trauma; thus, a previous Chiari malformation was considered [5,6].

Several symptoms are commonly observed in cases of syringomyelia, including segmental sensory loss (93%), pyramidal signs (82%), and muscle atrophy (60%) [3]. Irrespective of sensory or motor deficits, pain remains the predominant symptom, experienced by 50% to 90% of adult patients with syringomyelia. These pain symptoms may manifest as occipital or suboccipital headache, cervicalgia, or radicular pain [7]. Abnormalities in hindbrain cerebrospinal fluid (CSF) flow have been proposed as a pathogenic mechanism underlying headaches, particularly because symptoms can be exacerbated by the Valsalva maneuver. This correlates with the pathology associated with syringomyelia, which involves expansion of the "syrinx" containing cerebrospinal fluid that compresses the neurons of the lateral and anterior spinothalamic tracts. The posterior column is typically spared due to its more distal location. Consequently, patients often experience a loss of pain and temperature sensations with preservation of touch and vibration sensations, a phenomenon known as sensory dissociation.

In cases where vibration sensation is impaired, it is estimated that the syrinx is sufficiently enlarged to compress the posterior column. The distribution of sensory disturbances in such cases is often described as "cloak-like," predominantly affecting the shoulder region.
shoulder area) [3]. The clinical manifestations that appear depend on the size, location, and progressivity of the lesion.

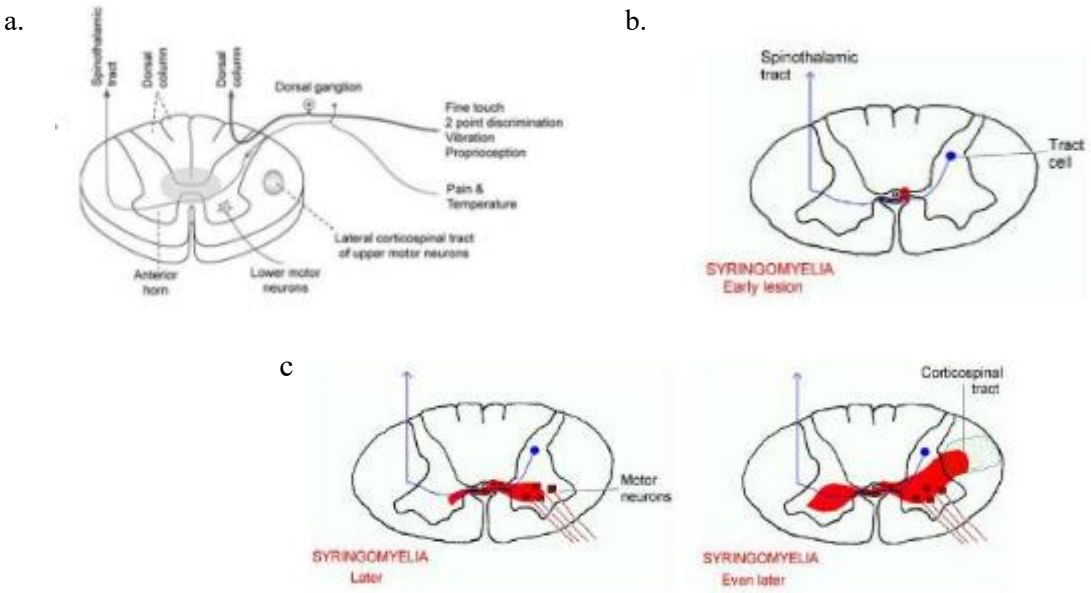

Figure 4. Lesson Progressivity of Syringomyelia

This begins when the syrinx expands and damages the motoneurons in the ventral horn, causing a lesion of the lower motoneurons at that location, for example, in the cervical spinal cord. Muscle wasting and weakness. In this patient, complaints first appeared 5 years ago; possibly the lesion had enlarged and touched the motor neurons. The motor neurons are unable to regulate signals to the muscles. This results in a loss of

muscle tone, or hypotonia, which can cause flaccidity and muscle atrophy. Preganglionic cell bodies of the sympathetic system exist in the lateral horn of the spinal grey matter. When the lesion affects this area, sympathetic disorders will appear, one of which is the sweat glands and manifests as hyper or anhidrosis.[9]

The MRI results revealed syringomyelia extending from the first cervical to the 11th thoracic vertebra, along with scoliosis. Initially, we discussed surgery options with the patient, but she ultimately decided not to proceed due to the associated risks. After consulting with rehabilitation specialists, she was advised to undergo cervical manipulation as the primary treatment. Several mechanisms could explain how this treatment may have reduced her symptoms, particularly pain. The goal is to prevent further spinal cord damage and enhance function, employing techniques described below. She may require active physical therapy, passive mobilization, and occupational therapy. We believe that cervical spine mobilizations might have helped alleviate the radicular aspects of her neck pain, aligning with a recent systematic review indicating that manual therapies can effectively reduce cervical radiculopathy [10] [11].

The patient received Infrared, Short Wave Diathermy (SWD) and Transcutaneous Electrical Nerve Stimulation (TENS) therapy and Physical Therapy for 1.5 years. The functions of these three modalities are for the physiological effects of heat therapy, including increasing cell metabolism, increasing the elasticity of connective tissue and muscles, increasing the excitability threshold and nerve conductivity, and vasodilatation of blood vessels. Nociceptive threshold, and acceleration enzyme activity. Non-surgical interventions were primarily physiotherapeutic and pharmacological therapeutic for 1.5 years, mostly by posture correction and exercises, and effects were alleviation of pain, improved physical function, improved activities of daily living, and quality of life. [10] [12] Forty-three of the patients managed surgically experienced clinical improvements, and 50% had symptom stabilization. Among the patients managed conservatively, symptom stabilization was attained in 88% and 2% improved clinically with symptom reduction. [10] A study conducted by Line et al shows that of 1,186 studies screened, seven studies met the eligibility criteria (4 single case studies and 3 cohort studies, a total of 90 individuals). The interventions were primarily physiotherapeutic, mostly by posture correction and exercises, and effects were alleviation of pain, improved physical function, improved activities of daily living, and quality of life. Syringomyelia patients should avoid activities that involve straining (e.g., lifting heavy objects, jumping) since these actions can trigger symptoms.[10] Medication therapies are betahistine 3x6mg, Noleptic 1x75mg, ranitidine 2x150mg, flamar gel, eperison 2x1, and celecoxib 1x100mg used for symptomatic complaints. The goals of conservative therapy in these patients are to prevent the progression of spinal cord damage, improve its function, and improve the quality of life of these patients. Patient felt better with the combination of conservative treatments above for just 1.5 years.

4. Conclusion

Syringomyelia is a complex neurological condition primarily affecting adults aged 20–50 years, often associated with Chiari I malformation, trauma, tumors, or inflammation. The pathogenesis involves hydrodynamic disruptions in cerebrospinal fluid flow, leading to syrinx formation and compression of spinal cord structures. Clinical manifestations include sensory dissociation, motor deficits, and pain, with symptoms varying based on syrinx size, location, and progression.

In this case, the patient presented with cervical to thoracic syringomyelia and scoliosis but declined surgical intervention. Instead, a conservative approach involving cervical manipulation, physiotherapy (Infrared, SWD, TENS), and pharmacological management was employed over 1.5 years, resulting in pain relief, functional improvement, and enhanced quality of life. Studies support that conservative management, particularly posture correction and exercise, can stabilize symptoms and improve daily functioning in many patients, though surgery remains an option for progressive cases. Ultimately, treatment should be individualized, balancing risks and benefits, with conservative therapies playing a significant role in symptom management and quality of life preservation for syringomyelia patients.

5. Data Availability Statement

The datasets generated and analyzed during the current study are not publicly available due to privacy and ethical considerations, but are available from the corresponding author upon reasonable request.

6. Ethical Statement

Sumatera Medical Journal (SUMEJ) is a peer-reviewed electronic international journal. This statement below clarifies the ethical behavior of all parties involved in the act of publishing an article in Sumatera Medical Journal (SUMEJ), including the authors, the chief editor, the Editorial Board, the peer-reviewer, and the

publisher (TALENTA Publisher Universitas Sumatera Utara). This statement is based on COPE's Best Practice Guidelines for Journal Editors.

7. Author Contributions

All authors contributed to the design and implementation of the research, data analysis, and finalizing the manuscript.

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10. Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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