Atypical Clinical Presentation of Syringomyelia: Review of Articles and a Case Report of Complete Unilateral Claw Hand Presentation

Siringomiyelinin Atipik Klinik Sunumu: Makalelerin Gözden Geçirilmesi ve Tam Tek Taraflı Pençe El Olgu Sunumu

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ABSTRACTS

Background: The typical presentation of syringomyelia is the combination of lower motor neuron signs at the segmental level, a dissociation sensory loss characterized by reduction of pain and temperature sensation with preservation of preserved light touch, vibration, and position sense in a cape pattern in the arms and upper trunk distribution. However, many clinical presentations do not conform to these typical features. The presentation may vary in the whole span of neurological signs and symptoms or can be non-specific.
Methods: To describe the clinical characteristic of reported atypical presentations of syringomyelia, its underlying cause and pathophysiology of its clinical presentations. **:** We reported a patient with syringomyelia presented with unilateral complete claw hand and case reports describing other atypical clinical presentations of syringomyelia published in the literature. We conducted a literature search on case reports and series describing atypical presentations of syringomyelia through PubMed/MEDLINE, Scopus, and Google Scholar databases. Demographic data, clinical characteristics, level of syringomyelia, underlying cause, surgical treatment, and outcome were extracted from selected article.
Results: A total of 23 case reports and case series were selected in this review based on the inclusion and exclusion criteria. The reported atypical clinical presentations could be themed into: 1) symptoms of peripheral neuropathy, 2) limb dystrophy, 3) limb hypertrophy, 4) movement disorder, 5) brainstem syndrome, 6) neuroarthropathy (NA), and 7) non-specific presentation.
Conclusion: Findings from this review article and case report would assist clinicians and physicians in the possibility of ruling in syringomyelia upon atypical clinical neurological presentation.

Key Words: Syringomyelia, atypical, clinical presentations, case reports

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ÖZET

Amaç: Siringomiyelinin tipik görünümü, segmental seviyedeki alt motor nöron belirtilerinin birleşimidir; kollarda pelerin şeklinde korunmuş hafif dokunma, titreşim ve pozisyon hissinin korunması ile ağrı ve sıcaklık hissinin azalması ile karakterize bir ayrışma duyusal kaybı ve üst gövde dağılımı. Bununla birlikte, birçok klinik sunum bu tipik özelliklere uymamaktadır. Sunum, nörolojik belirti ve semptomların tüm aralığında değişebilir veya spesifik olmayabilir.
Yöntem: Bildirilen atipik siringomiyelinin klinik özelliklerini, altta yatan nedenini ve klinik sunumlarının patofizyolojisini tanımlamak. : Tek taraflı tam pençe eli ile başvuran siringomiyelili bir hasta ve literatürde yayınlanan siringomiyelinin diğer atipik klinik sunumlarını anlatan vaka raporları sunduk. PubMed / MEDLINE, Scopus ve Google Scholar veritabanları aracılığıyla siringomiyelinin atipik sunumlarını açıklayan vaka raporları ve seriler üzerine bir literatür taraması gerçekleştirdik. Demografik veriler, klinik özellikler, siringomiyelinin düzeyi, altta yatan neden, cerrahi tedavi ve sonuç seçilen makaleden çıkarıldı.
Bulgular: Bu derlemede dahil etme ve dışlama kriterlerine göre toplam 23 vaka raporu ve vaka serisi seçilmiştir. Bildirilen atipik klinik sunumların temaları: 1) periferik nöropati semptomları, 2) ekstremite distrofisi, 3) ekstremite hipertrofisi, 4) hareket bozukluğu, 5) beyin sapı sendromu, 6) nöroartropati (NA) ve 7) spesifik olmayan sunum.
Sonuç: Bu derleme makalesi ve olgu sunumundan elde edilen bulgular, klinisyenlere ve hekimlere atipik klinik nörolojik prezentasyon üzerine siringomiyelide karar verme olasılığında yardımcı olacaktır.

Anahtar Sözcükler: Siringomiyeli, atipik, klinik sunum, olgu sunumları

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INTRODUCTION

The typical presentation of syringomyelia is the combination of lower motor neuron signs at the segmental level, a dissociation sensory loss characterized by reduction of pain and temperature sensation with preservation of preserved light touch, vibration, and position sense in a cape pattern in the arms and upper trunk distribution, and spinal long-tract upper motor neuron features below the level of the lesion (1). However, many clinical presentations do not conform to these typical features that are specific to the neuro-anatomical location of the syrinx. The presentation may vary in the whole span of neurological signs and symptoms or can be non-specific.

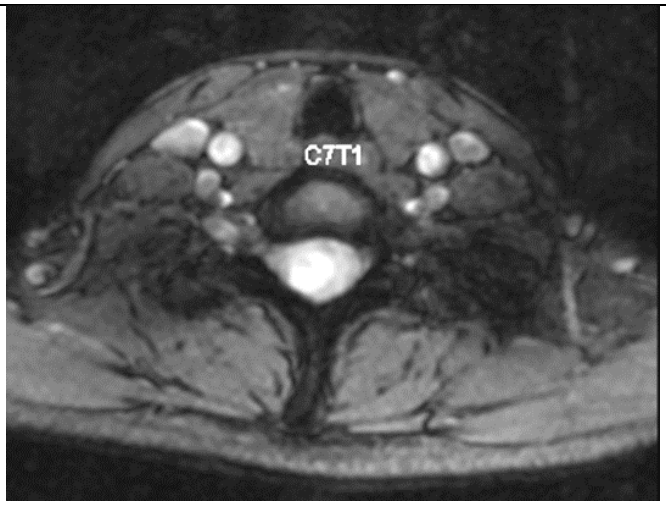
The impetus of this case report is to disseminate an atypical case of syringomyelia with unilateral complete claw hand presentation describing the etiology and mechanism of its development. A literature search and review of articles was conducted on established reported atypical clinical presentations of syringomyelia to illustrate the various features of clinical presentation, the possible mechanism behind these clinical signs and symptoms, and the outcome of the clinical conditions.

CASE REPORT

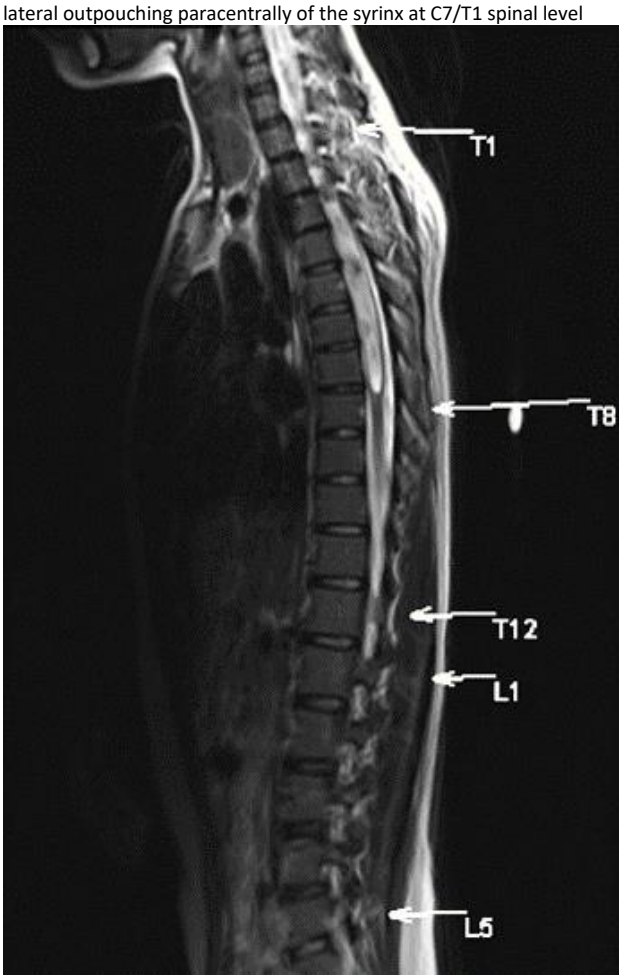
A 22-year-old lady, a right hand dominant, presented with a three months history of progressive unilateral right-hand weakness. It was accompanied by numbness that started distally and gradually progressed until the whole arm was involved. The weakness, however, was only confined to the distal area of the hand, slowly became prominent, leading to clawing deformity. Due to these symptoms, the fine motor function of her right hand was affected and hindering the performance of daily activities at home and work. The patient complained of having difficulty in manipulating small objects and equipment. These symptoms occurred in the absence of neuropathic pain, neurogenic bladder and bowel, or features that suggest increased intracranial pressure that would localize towards pathology in the central nervous system.

Clinical examination demonstrated the presence of marked atrophy of the thenar, hypothenar, and small intrinsic muscles of the right hand. It was associated with clawing of hand; notable hyperextension of the left metacarpal (MCP) joint, hyperflexion of left distal, and proximal interphalangeal (DIP and PIP) joint. The muscle strength of C5, C6, and C7 myotomes were all MRC (Medical Research Council) grade 5, while both C8 and T1 myotomes were MRC grade 2. There were marked weaknesses for thumb adduction, abduction, flexion, extension, and opposition movements, and reduced strength of flexor digitorum profundus and superficialis muscles, the intrinsic muscle of both palmar and dorsal interossei muscles. Each deep tendon reflexes of the right hand were hyporeflexia with an ipsilateral reduction in the pinprick sensation from C4 to T3 dermatomes. Proprioception was intact on both sides. Examination of the left upper limb and bilateral lower limb was unremarkable.

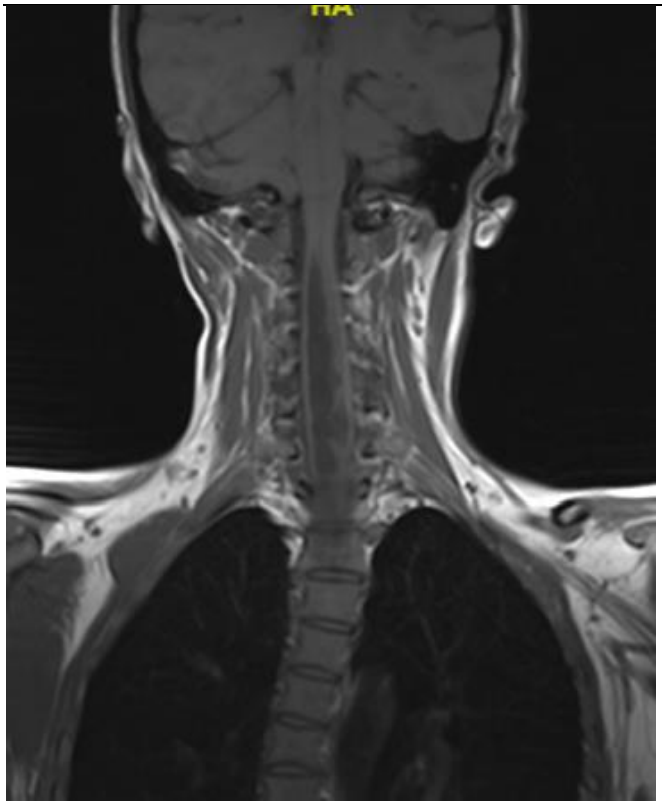
Nerve conduction study (NCS) demonstrated a marked reduction in the amplitude of compound muscle action potential (CMAP) in both the median and ulnar nerve with an otherwise normal finding of sensory nerve action potential (SNAP). Evaluation of CMAP and SNAP in the right radial nerve was normal. These findings were suggestive of a central cause warranting further imaging investigation of the brain and whole spine with MRI. The imaging revealed that the patient had a type 1 Arnold Chiari formation complicated with syringomyelia formation from C1 of the spinal cord down to the T8 level. Foramen magnum decompression surgery was done, and the patient had an uncomplicated postoperative recovery(Pictures 1-5).



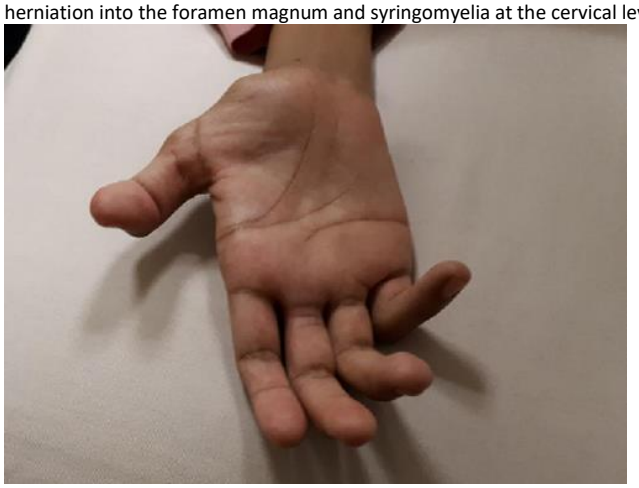
Picture 1: T2-weighted MRI cross sectional image demonstrates right posto-lateral outpouching paracentrally of the syrinx at C7/T1 spinal level



Picture 2: Sagittal plan T2-weighted MRI image demonstrates intramedullary signal abnormality of the spinal cord.



Picture 3: Frontal plane T1- weighted MRI image demonstrates the right tonsillar herniation into the foramen magnum and syringomyelia at the cervical level



Picture 4: Right thenar and hypothenar muscle wasting

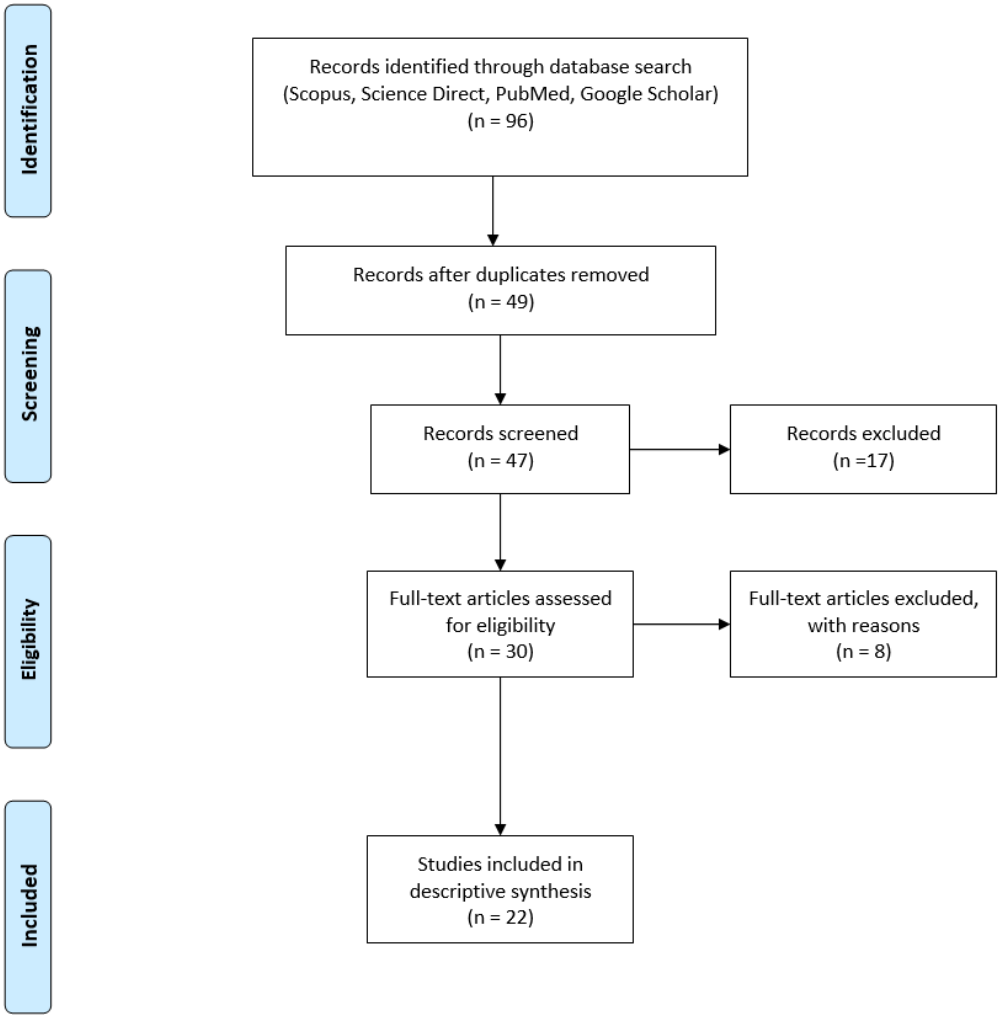
Picture 5: Right complete clawing of hand involving all digits

Method of Literature Search and Selection

Two authors conducted literature searches through PubMed/MEDLINE, Scopus, and Google Scholar bibliographic databases. Articles spanning from the year 1988 to 2019 were included. The keywords "syringomyelia", "hydromyelia", "atypical presentation", "atypical clinical" were searched in all possible combinations. Additional reference lists of all eligible studies were screened to be included.

Data Analysis

Inclusion criteria were: 1) presence of syringomyelia, 2) published in the English language, 3) case series and case reports, and 4) clinical presentation that is deemed atypical or unusual by the author(s). Exclusion criteria were: 1) clinical presentations with a complication of scoliosis, 2) post-traumatic spinal cord injury complicated with syringomyelia, 3) original studies, review articles and letter to editors, and 4) articles without full-text availability. Two authors independently screened the titles and abstracts of the articles gathered from the initial literature search. All authors arbitrated any discrepancies concerning the evaluation of the studies. A flow chart of the article selection is displayed in Figure 1. Data extracted include demographic (age and gender), characteristics of clinical presentation, level of syringomyelia, the underlying cause of syringomyelia, surgical treatment, and outcome.



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RESULTS

A total of 23 case reports and case series were selected in this review based on the inclusion and exclusion criteria.

The reported atypical clinical presentations could be themed into seven types: 1) symptoms of peripheral neuropathy, 2) limb dystrophy, 3) limb hypertrophy, 4) movement disorder, 5) brainstem syndrome, 6) neuroarthropathy (NA), and 7) non-specific clinical presentation. Descriptive data extraction from this literature is presented in Table 1.

Table 1: Article information summarizing atypical presentation of syringomyelia

Author(s)	Age / Gender	Atypical clinical Presentation	Level of syringomyelia	Cause of syrinx	Management	Outcome
Symptoms of Peripheral Neuropathy						
Pettorini et al (2)	13/F	Left claw hand	1st cervical vertebrae	CM1	Decompressive craniectomy, C1 laminectomy, opening of the dura and collagen matrix duraplasty	Good
Aly et al (3)	59/M	Ulnar Neuropathy secondary to elbow NA	Cervicothoracic	CM1	ND	ND
Scelsa et al (4)	24/M	Mimics ulnar neuropathy	Cervical	CM1	Sub occipital decompression with shunting of the syrinx to the cervical subarachnoid space	ND
Ziadeh et al (5)	26/F	Masquerading as Carpal tunnel syndromes	Cervico thoracic	CM1	Posterior decompression	Good
Jha et al(6)	20/F	9th and 10th cranial dysfunction (dysphagia and hoarseness of voice)	Medulla- cervical	Ependyoma	Laminectomy and tumour dissection	Good
Pilon et al(7)	30/F	Bilateral Abducens palsies and facial weakness	6th cervical vertebrae	CMI	Foramen magnum decompression	Good
Limb Dystrophy						
Sarder et al (8)	50/M	Muscles atrophy in upper back and shoulder girdle region	Cervico- medullary junction	CMI	ND	ND
Mora et al(9)	50/M	Distal muscle atrophy with bulbar dysfunction	Cervicothoracic	CM1	ND	ND
Sari et al (10)	17/M	Hemiatrophic Extremities, and Hypothalamic Hypogonadism	Cervical	CM1	ND	ND
Limb Hyperthrophy						
Pettorini et al (2)	9/M	Left arm hemihyperthrophy	Cervical	CMI	Suboccipital decompressive craniectomy, C1 laminectomy, opening of the dura and collagen matrix duraplasty	ND
Khanra et al(11)	32/M	Right Upper limb Hypertrophy sec to symphathetic overactivity	Cervicothoracic	ND	ND	ND
Movement Disorder						
Silva-Junior et al (12)	40/F	Cervical and axial dystonia	Cervicothoracic junction to the conus medullaris	CM1	Laminectomy with syringe -subarachnoid shunt.	Unchanged
Prasad et al (13)	4/F	Monolegia and Right torticollis	multiseptated syringomyelia involving the whole length of spinal cord from C1 down to the conus	CM1	Posterior decompression of the foramen magnum	Good
Berthet et al(14)	7/M	Movement disorder : Tics	Cervical	CM1	Posterior decompression and cervical laminectomy	Good
Hill et al (15)	29/M	Athethosis	Cervical to conus	Spina Bifida	revision of a blocked syringe-pleural shunt	Good
Hill et al (15)	48/F	Dystonia with retrocollis	Cervical	-	Symptomatic treatment with botulinum toxin injection and trihexyphenidyl	Poor
Brainstem syndrome						
Meves et al(16)	73/M	Brainstem syndrome : rotatory vertigo, nystagmus , dysarthria and dysphagia	Foramen magnum to thoracic	ND	ND	ND
Meves et al (16)	64/M	Brainstem syndrome : ataxic gait	Cervical	ND	ND	ND
Anwer et al (17)	38/M	Vertigo, facial weakness and Horner's syndrome	Cervical	CM1	Posterior fossa decompression	Good
Neuroarthopathy						
Nacir et al (18)		NA of multiple joint (Left shoulder , elbow , hand)	Cervicocothoracic	CM1	ND	ND
Singer et al (19)	72/F	Shoulder joint NA	Cervicothoracic	Unknown	ND	ND
Vemula et al (20)	55/M	Bilateral Ulnar neuropathy sec to elbow NA	cervicothoracic	CM1	Posterior fossa decompression with durapalsty	ND
Memarpour et al (21)	36/M	Hip Charcot NA	Cervical thoracic	CM1	Cervical decompression with a suboccipital craniectomy and C1 laminectomy	CM1
Wang et al(22)	52/F	Left shoulder joint	cervicothoracic	Basilar Impression	Posterior decompression	Good
Non- specific						
McManus et al(23)	19/M	Obstructive sleep apnoea	Cervical thoracic	CM1	Craniocervical decompressive procedure.	ND
Vanamoorthy et al (24)	34/M	Intractable hiccups	Cervical	CM1	Posterior fossa decompression	Good

*****CM1: Chiari Malformation Type 1, NA : Neuroarthopathy, ND : Not Disclosed, M : Male, F : Female*

DISCUSSION

Based on our review, Chiari Malformation is accounted for the most prominent underlying cause of syringomyelia with atypical clinical presentation. It is a congenital malformation of the brain stem, cerebellum, upper spinal cord, and surrounding bony structures, which consists of a series of congenital hindbrain herniation symptoms (1). The age of onset seen among 364 patients in a prospective cohort study was around 24.9+/-15.8 years, with a quarter of them cited trauma as the precipitating event. This condition is commonly associated with the formation of syringomyelia, with an incidence of 65% (25).

Presentation of syringomyelia can masquerade as peripheral neuropathy, which can be directly or indirectly caused by the syrinx. Claw hand presentation in Type 1 Chiari Malformation with syringomyelia is an uncommon presentation, as evident by very few similar published case reports (2-7). To the best of our knowledge, most of the reported syringomyelia-related cases with ulnar claw hand presentations were isolated and rarely conformed to the typical ulnar mononeuropathy feature. A case reported by Aly et al. (2) described a bilateral ulnar claw hand neuropathy due to ulnar nerve compression at the cubital fossa secondary to bilateral elbow neuroarthropathy. In contrast, another two reported cases on Type 1 Chiari Malformation presented with an isolated ulnar claw hand and T1 dermatome sensory loss, in which both were attributed directly to syringomyelia (2, 4). Ulnar neuropathy in these cases was ruled out with NCS and electromyogram (EMG) that was not conformed to peripheral ulnar neuropathy, hence the subsequent MRI investigation.

As reported in our case, the patient was presented with a unilateral complete claw hand feature affecting all the intrinsic muscles of the hand, including thenar and hypothenar. The mechanism of complete claw hand has resulted from paralysis of interossei and lumbrical muscles, leading to hyperextension of the MCP joint (26). Subsequently, the MCP joint is not stabilized, either at neutral or slight flexion position, blocking the effect of the transverse lamina on the long extensors tendon at this joint, thus reducing its ability to extend the interphalangeal joint. The enlargement of the syrinx can further explain this presentation as an asymmetrical localized paracentral out pouching at the level of the most significant axial cross-section, which often leads to local segmental signs of lower motor neuron lesion at the level of spinal involvement (25). Based on the axial T2 weighted MRI image of our patient, we have identified the presence of prominent asymmetrical paracentral outpouching of the syrinx towards the right at the C8/T1 spinal cord level. This finding was supported by the NCS that localised the lesion towards the cervical level. Our hypothetical explanation shares a similar case by Ziadeh et al. with a Carpal Tunnel Syndrome-like presentation evident by a syrinx with the most significant axial dimension at the C3-4 level, measured approximately 84mm (5).

Another cause of ulnar mononeuropathy is secondary to entrapment or compression by joint neuroarthropathy (3). Jha et al. reported the involvement of cranial nerve IX and X due to a sizeable intramedullary tumor in the cervico-dorsal region leading to a formation of syrinx that extended rostrally and compressed the nuclei of the cranial nerves (6). Pilon et al. illustrated similar pathological changes as these cranial nerves lie close to the exit of the pontomedullary junction. Hence they are susceptible to the compressive effects of intracranial malignancies, aneurysms, and cerebellomedullary malformations such as Type 1 Chiari Malformation (7).

Syringomyelia can present with dystrophic or atrophic changes of the muscles of the upper or lower limbs. As described by Mora et al., the case occurred at a young age with bilateral severe proximal muscle weakness and distal atrophic changes that led to the misdiagnosis of motor neuron disease (9). Type 1 Chiari Malformation with syringomyelia complication was diagnosed after NCS and EMG have demonstrated chronic neurogenic cause with denervation of C5 to T1, pointing towards spinal cord pathology. Sarder et al elucidated a similar case scenario with features of bilateral upper limb weakness, shoulder girdle muscle wasting and intact sensation masquerading motor neuron disease until NCS and EMG revealed unremarkable results (8). A hemiatrophic extremity is another atypical presentation reported by Sari et al. that accounts for widespread syringomyelia (10).

In contrast to limb dystrophic changes, a very unusual presentation of limb hypertrophy may need to consider syringomyelia (2, 11). The cause of the hypertrophy is still unknown, but it was stipulated that the hyperstimulation of preganglionic sympathetic neurons has resulted in hyperemia, thus the hypertrophic appearance (11). To objectively demonstrate the presence of an increased sympathetic tone, they performed a simple cold pressor test. The latter revealed an increase of vasomotor tone on affected hypertrophic limbs. Minor's test showed hyperhidrosis of the entire right upper limb, which further support the feasibility of increased sympathetic activity (11).

Various movement disorders, including dystonia, athetosis, torticollis, and tic, are very uncommon syringomyelia presentations, and the mechanism for these clinical presentations is yet to be understood (12-15). Nonetheless, many reported cases presented with movement disorder features improved with syrinx decompression (27). Electrophysiological findings suggest the role of damaged α-motor neuron, spinal interneurons lesion, and pathology at descending motor pathways as plausible contributing factors to movement disorders presentation (15).

Syringomyelia should be considered as one of the differential diagnoses in cases presenting with brainstem syndrome, especially with acute onset after ruling out vascular aetiology (16, 17). Symptoms have been reported to be in remission or triggered with certain positions or activities (16). This acute symptom may attribute to ischemia or mechanical distortion of the brainstem through an acute expansion of an existing cervical syrinx. Gardner et al. assumed that the intracranial arterial pulse wave is transmitted through a gentle water hammer effect and presses ventricular fluid through the central canal, initiating a cyst formation (28). Williams et al. proposed that the presence of an obstruction in the normal drainage of cerebrospinal fluid caused by the changes in the venous pressure leads to the enlargement of a pre-existing syrinx during postural changes and Valsalva maneuvers (29).

Acute neuroarthropathy (NA) appears to be an uncommon presentation of syringomyelia (18-21). A literature review by Wang et al. found a total of 17 cases presented as NA in the shoulder, 17 involving the elbow, 2 in the wrist and interphalangeal joints, and one over the hip and knee joint (22). Development of autonomic neuropathy stipulated by the neurovascular theory may suggest the presence of increased blood flow to the joint, the subsequent bone destruction and regeneration, and the following ultimatum of joint hypertrophy and architectural deformities (21).

Obstructive sleep apnoea can be a presenting complaint from syringomyelia. The anatomic localisation of respiratory centers in close proximity with the syrinx may explain the presence of respiratory disturbances (23). Another rather interesting presentation is intractable hiccup (23). The damage to the neurologic structures in the hiccup center by Chiari Malformation may be the likely mechanism (24).

The diagnosis of syringomyelia always comes much later, after ruling out other common etiologies. Accurate diagnosis is essential so that the right treatment can be administered. Most of the reported cases showed good outcomes with surgical intervention, as evident by the resolving clinical features. A systematic review evaluating the outcomes of four surgical techniques in managing syringomyelia has recommended a combination of bony decompression procedure with duroplasty to achieve a favorable outcome in reducing the symptoms and size of the syrinx (30). The overall improvement in clinical signs and symptoms of Type 1 Chiari Malformation with or without syringomyelia was 77% and 81%, respectively (30).

CONCLUSION

The clinical presentation of syringomyelia highly varies and does not always conform to the typical presentations of neuroanatomical lesions of the syrinx. We illustrated a few, if not all, the possible reported atypical clinical presentations that may guide clinicians towards the probable diagnosis of syringomyelia. Neurological deficits arise from syringomyelia are mostly reversible if the right management is administered. Hence, an early accurate diagnosis is imperative to limit the functional disabilities that would have resulted from misdiagnosing this condition.

Conflict of interest

No conflict of interest was declared by the authors.

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