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JCIMCR OPEN ACCESS Journal of Clinical Images and Medical Case Reports

ISSN 2766-7820

Case Report

Open Access, Volume 3

Sudomotor dysfunction and corneal nerve loss: An unusual presentation of syringomyelia and COVID-19 infection

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Received: Jun 27, 2022

Accepted: Jul 26, 2022

Published: Aug 02, 2022

Archived: www.jcimcr.org

DOI: www.doi.org/10.52768/2766-7820/1979

Abstract

Aim: To highlight unusual neurological findings in a patient with syringomyelia and COVID-19.

Methods: Neurological examination, Vibration Perception Threshold (VPT), Magnetic Resonance Imaging (MRI) of the full neuroaxis, electrophysiologic studies, Corneal Confocal Microscopy (CCM) and Sudoscan were performed.

Results: A previously fit and well 28-year-old lady presented with worsening shoulder and neck pain after COVID-19. She was found to have reduced pinprick sensation in both arms and base of the neck and MRI revealed a cervical syrinx. Electrophysiologic studies showed no evidence of upper limb neuropathy or cervical radiculopathy, but CCM showed corneal nerve loss and Sudoscan demonstrated severe sudomotor dysfunction in both hands.

Conclusion: We present an unusual case of worsening pain with sudomotor dysfunction and corneal nerve fibre loss in a patient with syringomyelia and COVID-19 infection.

Keywords: CCM; Sudomotor dysfunction; Syringomyelia.

Background and aims

Syringomyelia is characterized by cavitation within the spinal cord and typically presents with loss of pain and temperature sensation in a “cape-like” pattern with upper limb muscle atrophy and areflexia. COVID-19 infection may exacerbate painful diabetic neuropathy and has recently been associated with corneal nerve fibre damage. We have undertaken detailed neurological evaluation, MRI, nerve conduction studies, corneal confocal microscopy and evaluation of sudomotor function.

Case report

Clinical presentation

A 28-year-old female presented with a 2-year history of dull pain in both shoulders, upper back and neck which had worsened during pregnancy and after recent COVID-19 infection. The patient denied a history of trauma, weakness of the upper or lower limbs, visual disturbance or urinary or bowel dysfunction. There was no family or medical history of any other chronic disease or cause of neuropathy. Complete blood count, creatinine, HbA1c, ANA, B12 and folate were normal.

Neurological examination

There was a decrease in pinprick sensation in both upper extremities and base of the neck with a clear dissociation of temperature sensation over the back between her shoulders and mid-thorax in a typical ‘cape’ distribution. There was no muscle atrophy or fasciculation and upper and lower extremity muscles exhibited 5/5 strength based on the Medical Research Council Scale for muscle strength. Sensation to light touch, temperature, proprioception and reflexes were normal in both lower limbs.

MRI imaging

Magnetic Resonance Imaging (MRI) of the full neuroaxis was performed. The brain parenchyma was normal, diffusion weighted imaging excluded acute ischemic changes, and susceptibility imaging confirmed the absence of hemorrhagic disease. The cranio-cervical junction was at a normal level with no evidence of tonsillar ectopia or herniation. Spinal imaging showed normal vertebral bodies, and discs with no evidence of herniation. There was an intrinsic abnormality at C6-C7, which was hyperintense on T2 (Figure 1) without enhancement, consistent with the diagnosis of a cervical syrinx which was 3.4 cm in length with an AP diameter of 2.5 mm, without cord atrophy or expansion.

Neuropathy testing

The Vibration Perception Threshold (VPT) was normal in both upper and lower limbs (Table 1). Electrophysiologic studies revealed no evidence of large fibre neuropathy or cervical radiculopathy, with normal distal latencies, CMAP amplitudes and CVsin both median and ulnar motor nerves (Table 2). Corneal Confocal Microscopy (CCM) (Figure 2A,B) showed corneal nerve loss compared to an age-matched healthy control (Table 1). Sudoscan assessment demonstrated reduced Electrochemical Skin Conductance (ESC) consistent with sudomotor dysfunction in both hands with normal responses in the feet (Table 1).

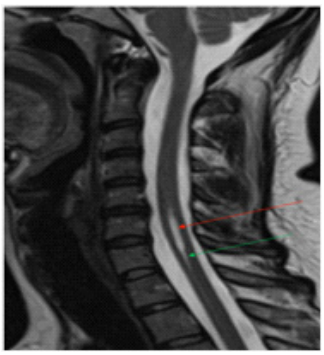

Figure 1: Sagittal T2 MRI of the cervical spine demonstrating a hyperintensity within the cord at C6-C7 level (red arrow). Note the "continuation" of signal from the central canal best demonstrated at the inferior margin (green arrow).

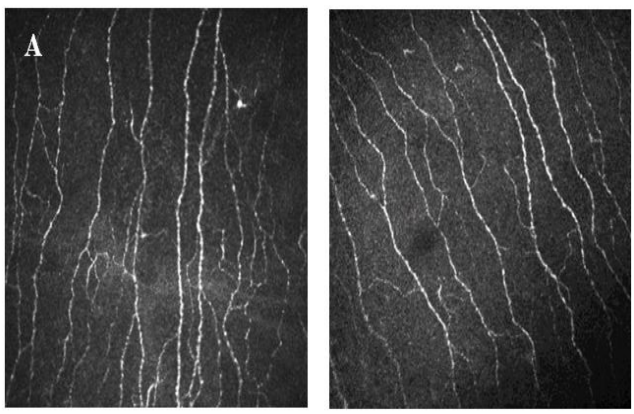

Figure 2: CCM image from a control subject (A) and the patient with syringomyelia (B).

Table 1: Corneal nerve, vibration perception and sudomotor function in the patient with syringomyelia and COVID-19 compared to an age-matched control.

	Patient	Control	Interpretation
Corneal nerve metrics			
CNFD (no./mm^2)	38.5	43.7	Moderate loss
CNBD (no./mm^2)	53.1	81.2	Moderate loss
CNFL (mm/mm^2)	22.4	25.8	Moderate loss
VPT			
	Patient	**Reference**	
R foot	5.0	≤ 15 V	Normal
L foot	3.5		Normal
Sudomotor function			
	Patient	**Reference**	
Feet	82	≥ 70 µS	Normal
Hands	39	≥ 60 µS	Severely reduced

CNFD: Corneal Nerve Fiber Density; CNBD: Corneal Nerve Branch Density; CNFL: Corneal Nerve Fiber Length; VPT: Vibration Perception Threshold; R: Right; L: Left; V: Volts; Ms: Micro Siemens.

Table 2: Upper limb nerve conduction studies in the patient with syringomyelia and COVID-19.

Nerve	Latency DML	Amplitude M-mV/S-uV	CV m/s	F-M Latency ms
Median motor left				
Wrist-APB	3.50	7.5	NS	20.8
Elbow-wrist	7.17	7.4	59.9	NS
Median motor right				
Wrist-APB	3.27	8.4	NS	20.7
Elbow-wrist	6.58	7.9	63.4	NS
Ulnar motor left				
Wrist-ADM	2.11	8.5	NS	21.4
Bl. Elbow-wrist	5.92	7.7	59.1	NS
Ulnar motor right				
Wrist-ADM	2.29	10.3	NS	23.8
Bl. Elbow-wrist	5.43	10.2	66.9	NS

DML: Distal Motor Latency; Mv: Millivolts; Uv: Microvolts; CV: Conduction Velocity; M/S: Meter Per Second; APB: Abductor Pollicis Brevis; ADM: Abductor Digiti Minimi; Ms: Milliseconds; NS: Not Stated.

Interpretation

Syringomyelia is associated with spinal cord cavitation, which typically affects motor and sensory tracts resulting in pyramidal and sensory deficits. Involvement of the spinothalamic tracts leads to a typical loss of pain and temperature perception in the upper limbs with pain, weakness and numbness. Focal dyshidrosis hasbeen reported in a case series of 30 patients with Chiari malformation and syringomyelia indicative of spinal cord sympathetic outflow damage in a distribution corresponding to the location of the syrinx [1].

Sudomotor dysfunction can be quantified using the Thermoregulatory Sweat Test (TST), Sympathetic Skin Response (SSR) and Quantitative Sudomotor Axon Reflex Test (QSART) and SudoscanTM, which measures Electrochemical Sweat Conduc-

tance (ESC). Recently a 22-year old male with MRI evidence of an Arnold Chiari malformation type 1 and syrinx cavity extending from the cervical cord to the cauda presented with excessive sweating on the left side of his body and Sudoscan confirmed a higher ESC on the left side (77 µS on left hand vs. 69 µS on right hand; 83 µS on left foot vs. 70 µS on right foot) [2]. Here we show selective sudomotor dysfunction in the hands with preserved responses in the feet and corneal nerve fibre loss. Indeed, we have recently shown loss of taste and smell with painful neuropathy and increased thermal thresholds in patients with diabetes [3] after acute COVID-19 and a loss of corneal nerves in individuals with long-COVID [4]. There is also a case series of patients with COVID-19 who developed autonomic symptoms and sudomotor and cardiovagal dysfunction [1]. This case serves to illustrate an atypical neurological presentation associated with syringomyelia and COVID-19.

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