Atypical clinical presentation of Syringomyelia: review of articles and a case report of complete unilateral claw hand presentation

ABSTRACT

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.

Keyword: Syringomyelia; Atypical; Clinical presentations; Case reports