Int J Clin Exp Med 2016;9(7):14568-14571
www.ijcem.com /ISSN:1940-5901/IJCEM0026199

Original Article

Syringomyelia in patient with neuromyelitis optica spectrum disorder: a case report

Min Tang, Qun Zhou, Youling Zhu

Department of Neurology, The Third Affiliated Hospital of Anhui Medical University, Hefei 230061, China

Received February 18, 2016; Accepted May 15, 2016; Epub July 15, 2016; Published July 30, 2016

Abstract: Neuromyelitis optica spectrum disorder (NMOSD) is an idiopathic inflammatory demyelinating disease characterized by longitudinally extensive transverse myelitis (LETM) and optic neuritis. This article reported a case of a 58-year-old women initially presented with asymmetric tetraparesis and binocular vision diminution. MRI (full name please) showed the longitudinal syringomyelia from C3 to T1 vertebral segments. Combined with the clinical manifestations and laboratory examinations especially seropositivity of NMO-IgG, the patient was ultimately diagnosed as NMOSD. This case report suggested that radiological findings of syringomyelia should be considered the possibility of NMOSD, especially in patients with a history of typical myelitis symptoms and seropositivity of NMO-IgG.

Keywords: Syringomyelia, neuromyelitis optica, neuromyelitis optica spectrum disorder, NMO-IgG

Introduction

Neuromyelitis optica (NMO, also known as Devic's disease) is a clinically defined, idiopathic inflammatory demyelinating disease of the central nervous system, characterized by optic neuritis and longitudinally extensive transverse myelitis (LETM) [1]. It is previously considered as a subtype of prototypic multiple sclerosis [2]. However, the detection of neuromyelitis specific antibody NMO-IgG has prompted revisions of the differential diagnosis of NMO as independent disease from other inflammatory diseases and broadened NMO to the new concept of neuromyelitis optica spectrum disorder (NMOSD) [1, 3, 4]. As the LETM has been listed as one of the major criteria for the diagnosis of NMOSD, it is important for us to be aware that spinal cord MRI showing continuous neuroimaging abnormalities must be considered the possible diagnosis of NMOSD [5]. The spinal cord MRI of NMOSD show the longitudinal long T2-weighted signal across the center of the spinal cord or spinal cord gray matter, long T1-weighted cord-like or patchy signal extending more than 3 vertebral segments, and significant spinal cord swelling in the acute phase.

Syringomyelia is a disorder in which fluid-filled cavities form inside the spinal cord which is triggered by intramedullary inflammation [6]. Previous studies demonstrated that syringomyelia cavitations, characterized by MRI lesion of hypointensity in T1-weighted images and hyperintensity in T2-weighted images, are a rare imaging feature in patients with NMO [7-9]. To our knowledge, there are no reports of syringomyelia in patients with NMOSD. The purpose of the article is to report a rare case represented as syringomyelia in spinal cord MRI and draw attention to this previously undefined feature of severe LETM in NMOSD.

Case report

Prior written and informed consent were obtained from the patient and the study was approved by the ethics review board of the Third Affiliated Hospital of Anhui Medical University. The patient is a 58-year-old woman who presented with progressively deterioration of occipital pain and asymmetric tetraparesis prominent on the right side lasting approximately one week. The tetraparesis mainly manifested as the inabilities of holding and standing in right upper and lower limb respectively. Her consciousness and excretory function were normal. Other symptoms including persistent logagnosia and hemianesthesia on the right side of the body especially the right lower extremity. Her

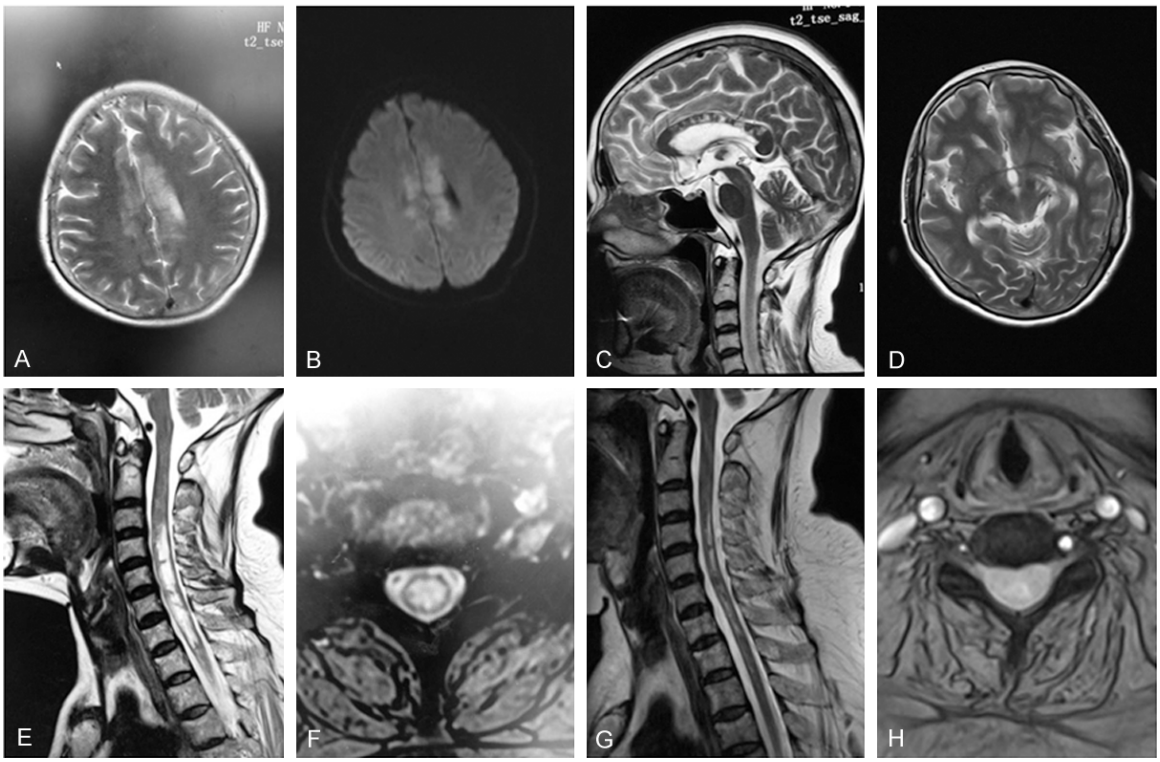

Figure 1. Brain and spinal cord MRI of the patient showing the infarction in corpus callosum and left centrum ovale (A-C), deformation changes in cerebral peduncle (D), and syringomyelia cavitations from C3 to T1 vertebral segments (E, F). After effective steroid therapies, the spinal cord MRI demonstrating the resolution of inflammatory lesions and syringomyelia cavitations (G, H).

history revealed that, before the onset of these symptoms, she was diagnosed as upper respiratory infection without fever, cough or expectoration; she had binocular vision diminution and then progressed into blindness in right eye in six months. Physical examination indicated that her time and spatial orientation was normal, pupils was normal with equal diameter of 3 mm, pupillary light reflex was poor in left-eye and absent in right eye, activity of eyeball activities was normal; ophthalmoscope examination showed binocular optic atrophy. Neurological examination suggested that both the facial sensation and pharyngeal reflex were normal; the deep and superficial sensibility of the fourth cervical nerve was attenuated at infraclavicular region; the muscle strength of right and left extremities were graded as 4 and 5 respectively; Examination of pathological reflex indicated that Lhermitte's sign (+), meningeal irritation sign (-), and bilateral Babinski sign (+). Laboratory test demonstrated that cerebrospinal fluid was clear and transparent, with pressure of 80 mmH_2O, Pandy test (+/-), nucleated cell 8.00×10^6/L, total protein 680 mg/L, glucose 4.10 mmol/L, chlorides 134.00 mmol/L, and IgG oligoclonal bands (-). Serum NMO-IgG (+). MRI of brain and spinal cord suggested that: (1) the infarction in corpus callosum and left centrum ovale(new onset) (**Figure 1A-C**); (2) the accumulation of cord-like abnormal signal in cerebral peduncle which was considered as deformation changes (**Figure 1D**); (3) syringomyelia from C3 to T1 vertebral segments (**Figure 1E**, **1F**). On the basis of these clinical and radiographic examinations, a diagnosis of NMOSD was confirmed. The patient was initiated with high-dose intravenous methylprednisolone pulse therapy (1000 mg daily for three consecutive days) and then switched to oral prednisolone (5 mg daily for 3 months). The spinal cord MRI after 3 months demonstrated that steroid therapies were effective in the resolution of inflammatory lesions and syringomyelia cavitations (**Figure 1G**, **1H**).

Discussion

The proposed concept of NMOSD reveals some special clinical cases undefined previously. The

diagnostic criteria for the definite NMO including the symptoms of optic neuritis and acute myelitis, and at least two of three supportive criteria including MRI demonstrating spinal cord lesions extending no less than 3 vertebral segments, MRI of brain lesions ruling out the diagnosis of multiple sclerosis, and seropositivity of NMO-IgG [10]. NMOSD have a wider sputum of clinical manifestation of brain lesions including LETM or optic neuritis in both of the proportion of patients with NMO and systemic autoimmune diseases [11]. We present one patient who developed syringomyelia in the spinal cord and brain. The particularity of this case is that, rather than spinal cord swelling, atrophic lesions of spinal cord and corpus callosum was prominent in MRI. Most notably, syringomyelia was the major performance of intramedullary signal. Combined with the more extensive lesions including cervical spinal and bilateral corpus callosum, and the history of binocular vision diminution, the diagnosis of relapsing NMOSD with complication of atypical spinal cord and brain lesions was considered for this patient.

Syringomyelia was also reported in patients with NMO in the previous studies [7-9]. In an earlier study, the imaging of extensive syringomyelia in the spinal cord s was displayed in 7 of 8 cases, which make authors assumed that these patients with syringomyelia may represented a new distinct entity [9]. After that, a Japanese study showed the gadolinium-enhanced dorsal syringomyelia from C1 to C7, suggesting an active demyelinating disorder in patient with NMO with positive NMO-IgG. And further study showed that syringomyelia disappeared after effective steroid medication for four years [8]. The most recent study from Turkey reported a case of a 10-year-old girl with longitudinal syringomyelia as a severe outcome of LETM from bulbus to T9 along the spinal cord [7]. Mechanism researches revealed that severe lesions of cervical spinal cord which could extend up to the medulla oblongata and induce a “presyrinx” state that ultimately lead to reversible development of syringomyelia [12].

The study indicated a serological positivity of NMO-IgG. NMO-IgG is a pathogenic antibody targeting the water channel aquaporin-4 and identifies NMOSD patients with high sensitivity of 72% and excellent specificity of 99.9% [4, 13]. Besides, the positive rate of NMO-IgG is higher in patients with NMO complicated by syringomyelia compared with that without syringomyelia [1]. Previous studies suggested that NMO-IgG significantly contribute to the pathogenesis of NMO and may facilitate the formation of syrinx through regulating water and glutamate homeostasis or enhancing blood-brain barrier permeability [14].

In summary, this report described a rare case of patient with NMOSD combined with syringomyelia from C3 to T1 vertebral segments. Due to the insufficient data, it’s too early to confirm the appearance of syringomyelia as a new clinical entity or an undefined feature of NMOSD. However, it indicates that radiological findings of syringomyelia should be interpreted cautiously and need to be considered the possibility of NMOSD, especially in patient with a history of typical symptoms and seropositivity of NMO-IgG. Finally, our case report serves as a warning for the future investigation of the profile of clinical and imaging characteristics in patients with NMOSD.

Acknowledgements

The authors thank Guangwu Li (MD, Prof. of medicine, Department of Neurobiology, Anhui Medical University) for kindly providing helpful advice in the conception, design and critical revision in this article.

Disclosure of conflict of interest

None.

Address correspondence to: Youling Zhu, Department of Neurology, The Third Affiliated Hospital of Anhui Medical University, Huaihe Road 390, Hefei 230061, Anhui Province, China. Tel: +86 0551621-83301; Fax: +86 055162183301; E-mail: okg333@126.com

References

[1] Nakamura M, Miyazawa I, Fujihara K, Nakashima I, Misu T, Watanabe S, Takahashi T and Itoyama Y. Preferential spinal central gray matter involvement in neuromyelitis optica. An MRI study. J Neurol 2008; 255: 163-170.

[2] Jarius S and Wildemann B. The history of neuromyelitis optica. J Neuroinflammation 2013; 10: 8.

[3] Kim W, Kim SH, Huh SY and Kim HJ. Brain abnormalities in neuromyelitis optica spectrum disorder. Mult Scler Int 2012; 2012: 735486.

[4] Lennon VA, Wingerchuk DM, Kryzer TJ, Pittock SJ, Lucchinetti CF, Fujihara K, Nakashima I and Weinshenker BG. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis. Lancet 2004; 364: 2106-2112.

[5] Jacob A, McKeon A, Nakashima I, Sato DK, Elsone L, Fujihara K and de Seze J. Current concept of neuromyelitis optica (NMO) and NMO spectrum disorders. J Neurol Neurosurg Psychiatry 2013; 84: 922-930.

[6] Ravaglia S, Bogdanov EI, Pichiecchio A, Bergamaschi R, Moglia A and Mikhaylov IM. Pathogenetic role of myelitis for syringomyelia. Clin Neurol Neurosurg 2007; 109: 541-546.

[7] Gokce G, Ceylan OM, Mutlu FM, Altinsoy HI and Koylu T. Relapsing Devic's disease in a child. J Pediatr Neurosci 2013; 8: 146-149.

[8] Sakabe E, Takizawa S, Ohnuki Y, Ohnuki Y, Kontani S and Takagi S. Syringomyelia in neuromyelitis optica seropositive for aquaporin-4 antibody. Intern Med 2010; 49: 353-354.

[9] Vernant JC, Cabre P, Smadja D, Merle H, Caubarrere I, Mikol J and Poser CM. Recurrent optic neuromyelitis with endocrinopathies: a new syndrome. Neurology 1997; 48: 58-64.

[10] Wingerchuk D, Lennon V, Pittock S, Lucchinetti C and Weinshenker B. Revised diagnostic criteria for neuromyelitis optica. Neurology 2006; 66: 1485-1489.

[11] Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ and Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol 2007; 6: 805-15.

[12] Fischbein NJ, Dillon WP, Cobbs C and Weinstein PR. The "presyrinx" state: a reversible myelopathic condition that may precede syringomyelia. AJNR Am J Neuroradiol 1999; 20: 7-20.

[13] Waters PJ, McKeon A, Leite MI, Rajasekharan S, Lennon VA, Villalobos A, Palace J, Mandrekar JN, Vincent A, Bar-Or A and Pittock SJ. Serologic diagnosis of NMO: a multicenter comparison of aquaporin-4-IgG assays. Neurology 2012; 78: 665-671; discussion 669.

[14] Hinson SR, McKeon A and Lennon VA. Neurological autoimmunity targeting aquaporin-4. Neuroscience 2010; 168: 1009-1018.