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Neuromyelitis optica spectrum disorder with spinal involvement including conus medullaris lesions with anti-aquaporin-4 antibody: A pediatric case and literature review

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Abstract

Introduction: Neuromyelitis Optica Spectrum Disorder (NMOSD) is an inflammatory disease of the central nervous system, generally associated with Anti-Aquaporin-4 (AQP4) antibody. It is characterized by severe attacks of optic Neuritis (ON) and Longitudinally Extensive Transverse Myelitis (LETM). Classically, it tends to spare the conus medullaris.

Case study: A 16-year-old girl was referred to our department for gait disturbance, numbness and heaviness of the right side of the body. Neurological examination revealed a right hemiparesis and posterior cord syndrome. Spinal MRI showed high signal lesions from T11 to L1, on T2/FLAIR weighted images with gadolinium enhancement. Immunological investigations revealed positivity of NMO-IgG/AQP4 antibody. The patient was treated with steroids and immunosuppressive drug with clinical improvement.

Conclusion: Our report intends to raise awareness of NMOSD in the pediatric population which may involve conus medullaris.

Keywords: Neuromyelitis optica spectrum disorder; Conus medullaris; Anti-aquaporin-4 antibody

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Introduction

Neuromyelitis Optica Spectrum Disorder (NMOSD) is a rare group of demyelinating syndrome of the central nervous system and an autoimmune astrocytopathic disease associated with Anti-Aquaporin-4 (AQP4) antibody [1]. Many studies have proven that NMOSD lesions have a tendency to be localized in the site of AQP4 expression such as spinal cord, white matter, hypothalamus, thalamus, third or fourth periventricular and midbrain [1,2]. The most common locations in spinal cord involvement are typically the cervical and thoracic ones [1]. Nonetheless, there are few reports on the conus medullaris lesions in NMOSD. Here we report the first pediatric case of a 16 year-old girl with anti AQP4 antibody positive NMOSD with conus medullaris involvement on Magnetic Resonance Imaging (MRI).

Case study

A 16-year-old Tunisian girl had medical personal history of corne alectasia and no family history of autoimmune disease. At the age of 15, she presented with numbness and heaviness in the right side of the body that spontaneously disappeared within a month. Neurological examination was normal. Brain MRI was normal. Spinal MRI showed right posterior and lateral cervical cord lesions (C3-C4) with no contrast enhancement. Eight months later, she presented with progressive numbness and heaviness on the same side with gait disturbance. Neurological examination revealed a right-sided hemiparesis predominant in the lower limb and posterior cord syndrome. The rest of the exam was normal including the absence of bowel, bladder or visual signs. Brain MRI was normal. Spinal MRI showed high signal lesions from T11 to L1, on T2/FLAIR weighted images with gadolinium enhancement (Figure 1).

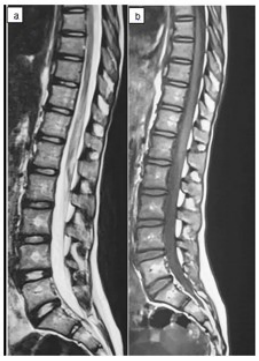

Figure 1: High signal lesions from T11 to L1, on T2 weighted images (a) with gadolinium enhancement (b).

Cerebrospinal Fluid (CSF) showed 10 el/mm^3 white cells, glucose level was 3.55 mmol/l (with corresponding serum glucose level of 3.7 mmol/l) and protein level of 0.2 g/l (normal <0.4g/l), no oligoclonal bands and IgG index was 0.63 (normal <0.65). Immunological investigations revealed positivity of NMO-IgG/AQP4 antibody. Cyanocobalamin level was normal. The diagnosis of NMOSD was made. The patient was treated by Intra Venous Methyl Prednisolone (IVMP) at the dose of 30 mg/kg/d for 5 days with significant improvement of motor signs. She had a persistent mild signs of right hemiparesis. The patient was discharged with oral prednisone at the dose of 1 mg/kg/d and azathioprine at the dose of 2 mg/kg/d. She had no longer posterior cordonal signs, and no deficit other than a mild pyramidal sign (right Babinski sign).

Discussion

We report the first pediatric case with conus medullaris involvement in NMOSD with positive AQP4 antibody. Only 9 cases were described in the literature (Table 1) [1,3-5]. The youngest case was 21 years-old reported by Sapnar et al [4].

Chitnis et al reported that the Wingerchuk 2006 criteria were 49% sensitive for a diagnosis of pediatric NMO, while the 2015 updated international panel for NMO diagnosis criteria were 97% sensitive and can be applied to children [6] .

An early diagnosis is mandatory to start treatment at an early stage and prevent relapses and disability progression in NMOSD. In acute attacks high doses of Intra Venous Methyl Prednisolone (IVMP) are recommended, as used in our patient. Moreover, recent studies have shown improved outcomes in patients treated with IVMP and plasma exchange compared to IVMP alone [2,7].

The maintenance treatment to avoid relapses includes oral and low-dose of steroids associated with a non-specific immunosuppressant drug (azathioprine, tacrolimus, mycophenolate-mofetil) [2,7,8] .

Conus lesions are overlooked as neuroimaging obtained typically in suspected NMOSD includes only the cervical and thoracic spinal cord [1].

To our knowledge, it is important to have in mind conus medullaris involvement possibility in children with AQP4 antibody positive NMOSD, therefore, lumbosacral MRI may be significantly valuable for early diagnosis and treatment.

Table 1: Reported cases with AQP4-antibody positive NMOSD with conus medullaris involvement.

Ref	Age (y)/ Gender	Clinical features	Age of onset (y)	MRI	Treatment	Evolution
[1]	25/F	Nausea, vomiting, progressive numbness in the lower extremity, progressive bilateral visual loss, dysuria, incontinence, loss of consciousness, absent joint position and vibration sensations.	25	Right frontal, temporal lobe, hippocampus lesions, without gadolinium enhancement; C5-C6 cervical vertebra, and T2-L1 thoracic vertebra lesions.	UK	UK
[2]	47/F	Progressive weakness of lower limbs and gait disturbance. Hypoesthesia below the C6 vertebra, absent joint position and vibration sensations Abdominal pain with belt-shaped numbness, urinary and fecal incontinence.	47	Bilateral frontal lesions, without gadolinium enhancement; high signal lesions over C2-3, C6-8 and L1-3 on T2-weighted images.	UK	UK

[3]	31/F	Gaitdisturbancewith progressive leftlegweakness and numbness.	31	Extension of abnormal T2-weighted signal within the cervical 1–2, 4–6, thoracic 6–7, 9–10, and 11–12 level spinal cord and medulla. Multifocal nodular T2 high-signal lesions in both Periventricular and subcortical white matter.	Steroid pulse therapy. low dose steroid and azathioprine for secondary prevention.	A slight improvement of left leg weakness. Four months later, right lower extremity numbness.
[4]	21/F	Area Postrema syndrome; later, urinary incontinence and saddle anesthesia.	21	Lesion in the conus medullaris and contrast enhancement of the nerve roots in the cauda equina.	Plasmapharesis.	Near complete resolution of symptoms.
[5]	79/F	Gait disturbance, right leg weakness, vibratory sense disturbance in both legs, hyporeflexia in the right achilles tendon.	72	T2 high-intense lesion with gadolinium enhancement at conus medullaris. Enhancement in spinal roots and cauda equina.	2 courses of IV methylprednisolone. Plasma exchanges (4 times), then low doses of prednisolone.	Improvement after plasma exchanges. No relapses.
[6]	67/F	Rapid progression of muscle weakness of both legs. flaccid paraparesis, sensory disturbance below L2 level, areflexia of lower extremities, dysuria, and constipation.	67	Diffuse high-intense Lesion extending from Th11 to the conus medullaris on T2-weighted image.	6 plasma exchanges No immunosuppressive treatment.	Good recovery. No exacerbations
	UK	UK	UK	extensive longitudinally transverse myelitis from cervical or upper thoracic levels to conus medullaris	UK	UK
	UK	UK	UK		UK	UK
	UK	UK	UK	isolated conus lesion	UK	UK

References

1. Wang Y, Zhang Y, Shan Y, Lu Z. Two Cases of Conus Medullaris Lesions with Neuromyelitis Optica Spectrum Disorders. J Mult Scler (Foster City). 2016; 3: 164.
2. Bradshaw MJ, Vu N, Hunley TE, Chitnis T. Child Neurology: Neuromyelitis optica spectrum disorders. Neurology. 2017; 88: e10-e13.
3. Kim S, Kwon B, Park J, Lee H, Kim H, et al. Neuromyelitis Optica Masquerading as Lumbosacral Radiculopathy: A Case Report. Ann Rehabil Med. 2016; 40: 943-948.
4. Sapnar S, Prabhu A. Lumbosacral Radiculomyelopathy-a rare presentation in an established case of Neuromyelitis Optica Spectrum Disorders. Neurology. 2017; 88.
5. Takai Y, Misu T, Nakashima I, Takahashi T, Itoyama Y, et al. Two cases of lumbosacral myeloradiculitis with anti-aquaporin-4 antibody. Neurology. 2012: 79: 1826-1828.
6. Chitnis T, Ness J, Krupp L, Waubant E, Hunt T, et al. Clinical features of neuromyelitis optica in children US Network of Pediatric MS Centers report. Neurology. 2016; 86: 245-252.
7. Abboud H, Petrak A, Mealy M, Sasidharan S, Siddique L, et al. Treatment of acute relapses in neuromyelitis optica: Steroids alone versus steroids plus plasma exchange. Mult Scler. 2016; 22: 185-192.
8. Kleiter I, Gahlen A, Borisow N, Fischer K, Wernecke KD, et al. Neuromyelitis optica: evaluation of 871 attacks and 1,153 treatment courses. Ann Neurol. 2016; 79: 206-216.