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*Clinical Case*

# First documented case of Devic's neuromyelitis optica in Niger

## Premier cas documenté de neuromyélite optique de Devic au Niger

H. Assadeck[1], M. Toudou Daouda[2], Y. Maiga[3], F. Hassane Djibo[2], D. Douma Maiga[1], E. Adehossi Omar[1,2]

**Abstract**

Devic's neuromyelitis optica is a chronic inflammatory demyelinating disease of the central nervous system that mainly affects spinal cord, optic nerve and brain regions with high aquaporin 4 antigen expression. We report the first documented case of Devic's neuromyelitis optica in Niger. It was a 66-year-old black man who had presented a rapidly progressive flaccid tetraplegia associated with vesico-sphincteral disorders, in whom magnetic resonance imaging had shown longitudinally extensive transverse cervical myelitis with positive anti-NMO antibodies.

***Keywords:*** Tetraplegia, Magnetic resonance imaging, Anti-NMO antibodies, Devic's neuromyelitis optica, Niger

**Résumé (French summary)**

La neuromyélite optique de Devic est une pathologie inflammatoire chronique démyélinisante du système nerveux central qui affecte électivement la moelle spinale, le nerf optique et les régions cérébrales à haute expression d'antigènes aquaporine 4. Nous rapportons le premier cas documenté de neuromyélite optique de Devic au Niger. Il s'agissait d'un homme de 66 ans de race noire qui avait présenté une tétraplégie flasque d'installation rapidement progressive associée à des troubles vésicosphinctériens, chez qui l'imagerie par résonance magnétique médullaire avait montré une myélite cervicale transverse longitudinalement étendue avec des anticorps anti-NMO positifs.

***Mots-clés:*** Tétraplégie, Imagerie par résonance magnétique, Anticorps anti-NMO, Neuromyélite optique de Devic, Niger

## Introduction

First described in 1894 by *Eugene Devic* [1] and *Fernand Gault* [2], Devic's neuromyelitis optica (NMO) or Devic's disease is a severe chronic inflammatory demyelinating disease of the central nervous system (CNS) that mainly affects spinal cord, optic nerve and brain regions with high aquaporin 4 antigen expression. In sub-Saharan Africa, very few studies and case series on Devic's disease have been reported in the literature **[3 – 6]**.

The incidence of NMO in sub-Saharan Africa is difficult to assess due to the rarity of studies on this disease in sub-Saharan populations. We report the first documented case of Devic's neuromyelitis optica in Niger.

**Clinical case**

A 66-year-old black man from Niger with no known past medical history, consulted in May 2017 for rapidly progressive limb weakness associated with vesico-sphincteral disorders type of urinary incontinence that occurred 1 month before consultation. He reported no history of trauma of the spine, or fever or exposure to chemical or toxic products. On admission, neurological examination found an areflexic quadriplegia predominantly in the lower limbs, associated with sensory level D3. He was afebrile, hemodynamically stable and with normal respiration. He did not report visual disturbances since the onset of symptoms. He does not report recent or previous visual disturbances as well as sensory and motor disorders.

Spinal-cord magnetic resonance imaging (MRI) showed a transverse cervical myelitis, hyperintense on T2-weighted images and extending from C6 to D3, and isointense on T1-weighted images, with enhancement on gadolinium administration **(*Figure 1*).** His brain MRI shows a non-specific left parietal cortical lesion **(*Figure 2*).**

Laboratory examinations (urea, creatinine, glycemia, blood count, C reactive protein, serum electrolytes, erythrocyte sedimentation rate) was normal. A cerebrospinal fluid (CSF) study revealed a white blood cell count of 3/m3, with glucose at 0.42 g/L and protein at 0.25 g/L, without oligoclonal IgG bands. Immunological tests revealed positive anti-neuromyelitis optica (anti-NMO) antibodies by using indirect immunofluorescence of transfected cells, with positive antinuclear antibodies. Anti-double-stranded DNA (anti-dsDNA) antibodies, anti-Smith (anti-Sm), anti-Sjögren's syndrome types A and B (anti-SSA and anti-SSB) and antiphospholipid antibodies, as well as the dosage of angiotensin-converting enzyme (ACE), were all normal. Serological tests for syphilis, human immunodeficiency virus (HIV), hepatitis C and hepatitis B, cytomegalovirus (CMV), the Epstein–Barr virus (EBV), herpes simplex virus (HSV) 1 and 2, were also negative. The thyroid function tests (anti-thyroperoxydase antibodies, anti-thyroglobulin, TSH, T3 and T4) were normal. The visual potentials evoked were normal. The search for pulmonary and extrapulmonary tuberculosis (Chest X-ray, mycobacterium tuberculosis) was negative.

At the end of these analyses, a diagnosis of Devic's neuromyelitis optica was considered. The patient was treated with pulse intravenous methylprednisolone (1 g/day for 10 days), relayed by 3-months oral corticotherapy (1 mg/kg/day) with azathioprine (150 mg/day) in the long-term.

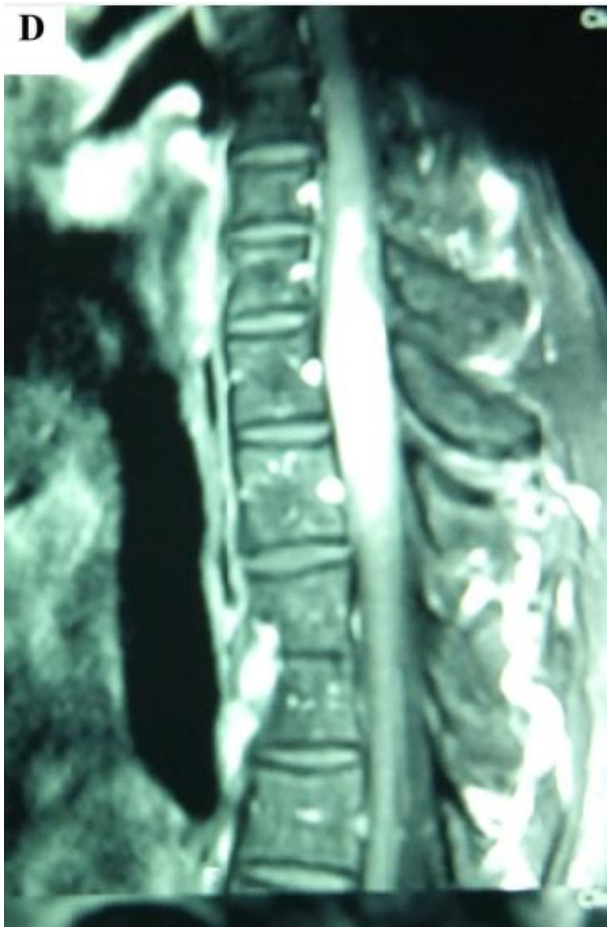


**Figure 1 :** Spinal-cord showing a transverse cervical myelitis, hyperintense on T2-weighted images (A, C) and extending from C6 to D3, and isointense on T1-weighted images (B), with enhancement on gadolinium administration (D).

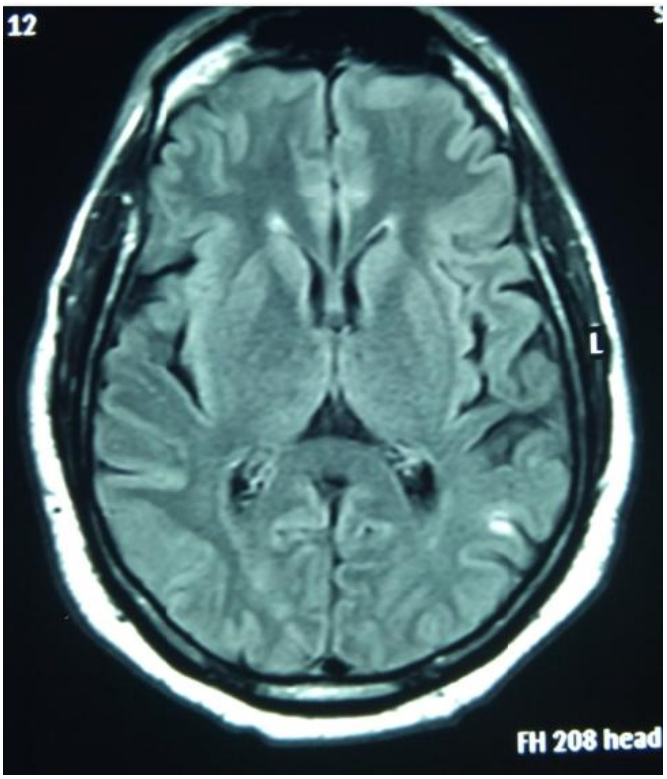


**Figure 2 :** Cerebral MRI showing a left parietal cortical lesion on FLAIR sequence.

**Discussion**

Our case report describes the first documented case of Devic's neuromyelitis optica in Niger. The diagnosis of NMO was considered in our patient after ruling out other etiologies which can give longitudinally extensive transverse myelitis, and further confirmed by the positivity of anti-NMO antibodies.

The age at onset of symptoms in our patient was 66 years, which is a very late start age compared to the data of previous studies in sub-Saharan Africa, in which the mean age at onset of symptoms ranged from 29 to 40 years [3 – 7]. In South Africa, NMO affects more women than men, with a sex ratio female/male ranging between 3 and 3.7 [4, 6]. This female predominance was reported in Senegal by *Dadah et al.* in 2017 [7]. However, in 2016 in

Mali *Maiga et al.* [5] did not report a prevalence of sex in their small case series of NMO.

The diagnosis of NMO in sub-Saharan Africa remains a diagnostic challenge in some countries for practitioners due to limited access to diagnostic tools. In these countries, the diagnosis is mostly clinical but in some cases MRI (when it is performed) can show characteristic lesions of NMO. These diagnostic difficulties make the Devic's disease relatively rare in sub-Saharan populations, because it is underdiagnosed. The recent availability of MRI and immunological tests in some sub-Saharan African countries will help in the disease's diagnosis and follow-up in most cases.

## Conclusion

NMO seems to be rare in Niger and often a diagnostic challenge due to limited access to MRI and lab tests such as anti-NMO antibodies or anti-MOG (anti-myelin oligodendrocyte glycoprotein). A multicentric study is needed in sub-Saharan Africa to gain better knowledge of this disease in sub-Saharan populations.

*** Corresponding author:**
Hamid Assadeck (assadeckhamid@yahoo.fr)

[1]Department of Medicine and Medical Specialties, Faculty of Health Sciences, Abdou Moumouni University of Niamey, Niger

[2]Department of Internal Medicine, National Hospital of Niamey, Niger

[3] Department of Neurology, Gabriel Touré University Teaching Hospital of Bamako, Mali

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