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Case Report

Diagnosis of a Rare Case of Acute Transverse Myelitis

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Abstract

Acute transverse myelitis is an acute inflammatory disease of the spinal cord characterized by rapid onset of bilateral neurological symptoms. Weakness, sensory disturbance, and autonomic dysfunction evolve over hours or days progressing to maximal clinical severity within 10 days of onset. It remains a rare neurological condition. We report the case of a 19-year-old man who presented with bilateral neurological disorders of progressive onset. MRI of the spinal cord showed inflammatory involvement of the cord. In correlation with the rest of the etiological workup, the diagnosis of ATM was retained. The treatment was corticosteroid therapy with good and progressive evolution. Neurological sequelae also occurred. Several differential diagnoses exist, it's essentially about other inflammatory or non-inflammatory myelopathies which can lead to wandering in the etiological treatment.

Keywords: *Acute Myelitis; Idiopathic; MRI; Thoracic Spinal Cord; Demyelination*

Abbreviations

ATM: Acute Transverse Myelitis; MRI: Magnetic Resonance Imaging; CSF: Cerebro Spinal Fluid; TMCWG: Transverse Myelitis Consortium Working Group

Introduction

Acute transverse myelitis (ATM) is a rare neurological disease secondary to inflammation of the spinal cord within 4 hours to 21 days from the onset of symptoms resulting in a syndrome of sensory-motor and autonomic neurological manifestations. It results from a dysfunction of sensory, motor, and autonomic functions secondary in most cases to a pathology. It is classified as idiopathic according to very precise criteria in the absence of a proven etiology. Epidemiologically, its incidence varies from 1 to 5 cases per million inhabitants per year. There is no notion of family heredity or gender preference [1]. Its definition has evolved over time with a consensus through the criteria of the Transverse Myelitis Consortium Working Group. Diagnosis is made clinically through neurological symptoms of sudden or progressive onset. The patient's history and associated signs play an important role in the diagnosis. The imaging of choice for the detection of abnormalities remains MRI. It will show abnormalities of the spinal cord. Treatment will depend on the etiology. The use of corticosteroids and other adjuvant treatments is appropriate. Rehabilitation will be given to patients. Neurological sequelae may be irreversible.

Case Report

We report the case of 19-year-old man presenting with acute onset of bilateral progressive sensory deficit, paraparesis, bowel and urinary dysfunction. Symptomology has been evolving for two weeks. The patient had no history of ophthalmologic, cardiac or pulmonary pathology, nor of parasitosis. At admission the neurological examination revealed a bilateral well-defined sensory level below T1 and paraplegia.

An MRI was performed to look for spinal cord abnormalities. Cervical spinal MRI shows on sagittal T2-weighted image through the thoracic spine demonstrates cord expansion and T2 hyperintense signal extending slightly over two spinal segments (Figure 1). Axial- T2 weighted image shows diffuse increased cord signal (> 2/3 of cross-sectional area of cord) (Figure 2).

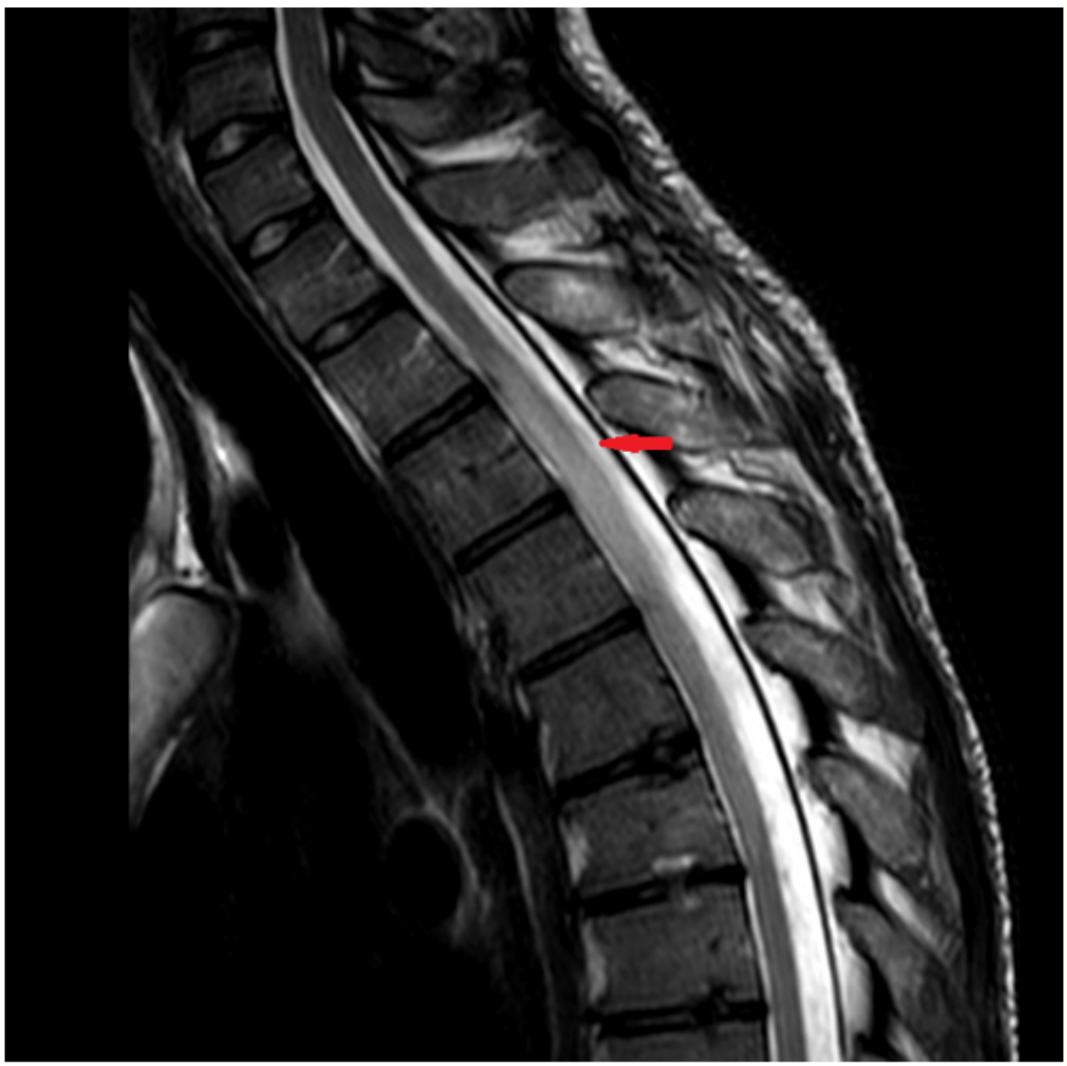

Figure 1: *Sagittal T2-weighted image through the cervico-thoracic spine demonstrates cord expansion and T2 hyperintense signal extending slightly over two spinal segments*

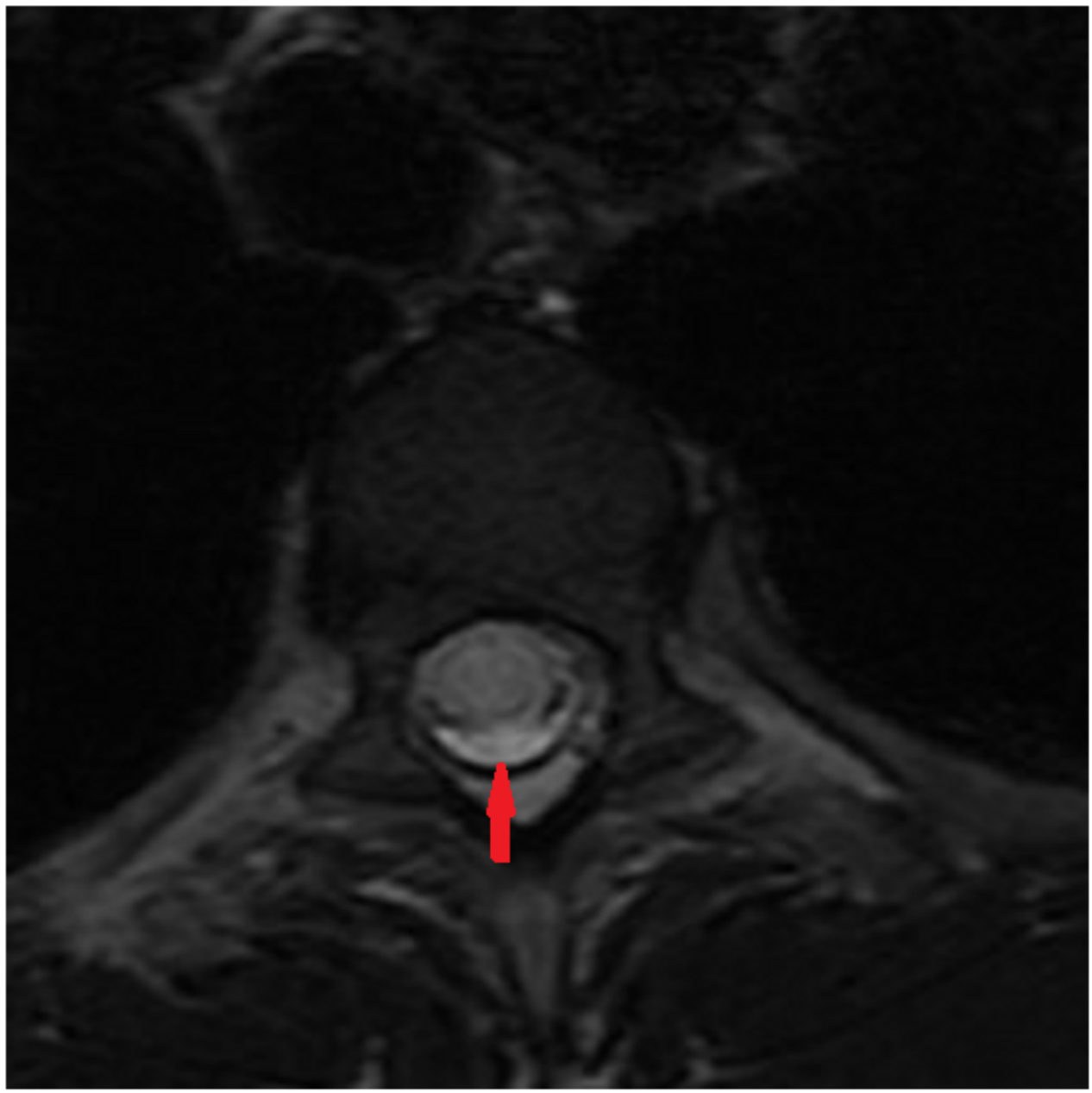

Figure 2: *Axial- T2 weighted image shows diffuse increased cord signal (> 2/3 of cross-sectional area of cord).*

On Contrast enhanced sagittal T1-weighted image with fat suppression through cervicothoracic spine reveals focal enhancement within the abnormal thoracic spinal cord segment that is much smaller than the extent of T2 signal abnormality (Figure 3). There is no sign of medullar compression on the spinal MRI.

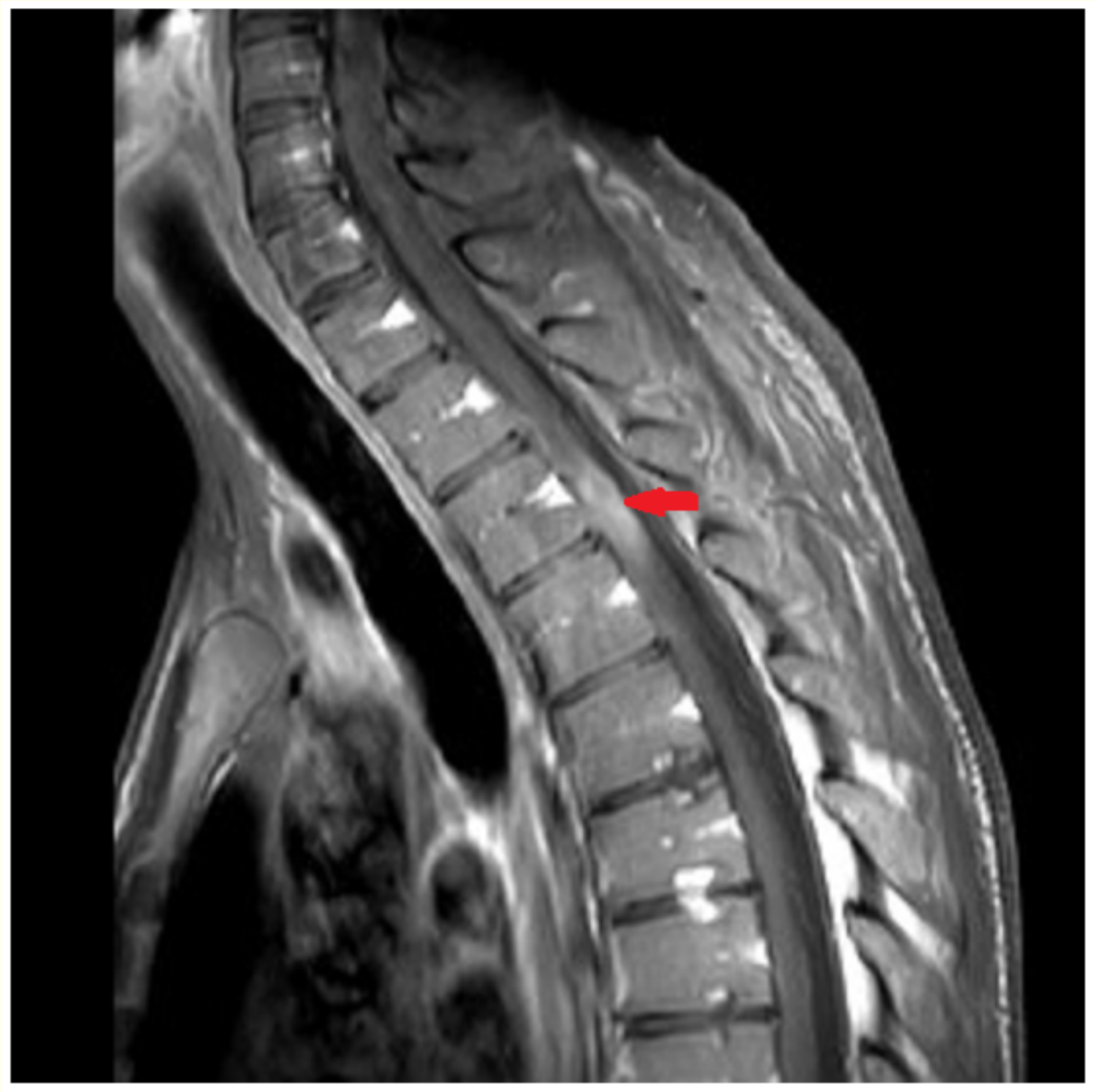

Figure 3: *Contrast enhanced sagittal T1-weighted image with fat suppression through cervico-thoracic spine reveals focal enhancement within the abnormal thoracic spinal cord segment that is much smaller than the extent of T2 signal abnormality.*

Brain MRI was normal.

Blood tests were normal.

Cerebro Spinal Fluid (CSF) findings show CSF pleocytosis, elevated IgG index and leukocytosis.

The search for anti-NMO antibodies was negative, as were the serological tests (HIV, Syphilis, Toxoplasmosis, Borreliosis).

For the purpose of etiological research, a thoraco-abdomino-pelvic CT scan was also carried out but did not provide any information.

The final diagnosis was acute transverse myelitis idiopathic according to the Transverse Myelitis Consortium Working Group (TMCWG).

The patient was treated with corticosteroids with a marked improvement in his condition in the short term. An immunosuppressive treatment was associated. Functional rehabilitation was also started.

Discussion

ATM is an acute inflammatory disease of the spinal cord characterized by rapid onset of bilateral neurological symptoms. Weakness, sensory disturbance and autonomic dysfunction evolves over hours or days progressing to maximal clinical severity within 10 days of onset [2].

It remains a rare neurological condition with a peak between the two decades of 10-19 years and 30-39 years [1]. E Smith., *et al*. in northern Finland in a retrospective study from 1 January 2002 to 31 December 2010, found a frequency of 1.04 cases/100,000 inhabitants/year, according to the TMCWG criteria, with only 63 patients diagnosed with ATM and 7% is idiopathic. The age range was 35 +/- 14.9 years with 62% women [2]. Zalewski., *et al*. in 5-years study, found 18% ATM, a median age of 48 years (18 - 79) and 61.9% women [3]. In his study, West., *et al*. found an incidence of 24.6 per million with an idiopathic ATM rate of 6.2 per million [4].

The symptomatology is progressive and dominated by lumbar pain, myalgias, bilateral neuro sensitive motor disorders, ranging from paresthesia of the lower limbs to paraplegia. Depending on the stage, it will be associated with disorders of the autonomic system, notably sphincter disorders, thermoregulation disorders (sweating), and tension disorders, all of which evolve within 10 days [1,5].

The diagnosis meets the criteria of the Transverse Myelitis Consortium Working Group (TMCWG): Sensory, motor, or dysautonomic involvement of spinal cord origin. Bilateral symptoms (not necessarily symmetrical) defining a sensory level [1,6] (Table 1):

- Sensory, motor or autonomic dysfunction attributable to spinal cord.
- Bilateral signs and symptoms.
- Well-defined sensory level.
- Cord inflammation confirmed by CSF pleocytosis, elevated IgG index or gadolinium enhancement.
- Progression to nadir between 4 hours and 21 days from onset.
- Symptoms must progress if present upon awakening.
- Compressive etiology excluded by neuroimaging.
- May repeat MRI ± lumbar puncture between 2 - 7 days if signs of inflammation absent initially.

Other clinical criteria were suggested in 2007 by TF Scott., *et al*. to complement this consensus definition [7,8]:

Diagnostic criteria for transverse myelitis
Inclusion criteria for diagnosis of transverse myelitis (idiopathic or disease associated)
Development of sensory, motor or autonomic dysfunction attributable to the spinal cord
Bilateral symptoms
Clearly defined sensory level
Exclusion of compressive aetiology by MRI or CT myelography
Spinal cord inflammation demonstrated by CSF pleocytosis or elevated IgG or gadolinium enhancement
Progression to clinical nadir between 4 hours and 21 days from onset of symptoms
Exclusion criteria for diagnosis of transverse myelitis (idiopathic or disease associated)
History of radiation to the spine within 10 years
Clear arterial distribution clinical defect consistent with anterior spinal artery occlusion
Abnormal flow voids on the surface of the cord consistent with AVM
Exclusion criteria for idiopathic transverse myelitis
Serologic or clinical evidence of connective tissue disease (sarcoidosis, Behcet's disease, Sjogren's syndrome, SLE, mixed connective tissue disorder)
CNS manifestations of syphilis, Lyme disease, HIV, HTLV-1, Mycoplasma, other viral infection.
Brain abnormalities suggestive of MS
History of clinically apparent optic neuritis

Table 1: *Diagnostic criteria for transverse myelitis of transverse myelitis consortium working group, 2002.*

- Complete transverse myelitis: Moderate to severe symmetrical motor involvement and dysautonomia of spinal cord origin. Presence of a sensory level.
- Partial transverse myelitis: Mild sensory and/or motor involvement, bilateral or unilateral, of spinal cord origin. When a severe deficit is present, a marked asymmetry is observed. Symptoms defining a bilateral or unilateral sensory level or presence of a typical myelitis MRI.

The gold standard in diagnosis remains spinal cord MRI, which will highlight abnormalities in the spinal cord. It will allow the exploration of the entire spinal cord to determine the extent of the lesions and eliminate the causes of compression. It will be associated with a cerebral MRI.

Although any cord level can be affected with a preference for the thoracic cord. Imaging can be normal in up to 40% of cases.

MRI findings:

- Cord expansion in the acute phase extending over more than two segments in the sagittal plane and involving more than two-thirds of the cross-sectional area of the cord.
- Abnormal central cord signal that is iso-hypointense on T1-WI and hyperintense on T2-WI/STIR.
- Enhancement patterns are variable: diffuse enhancement, heterogeneous enhancement, and peripheral enhancement has all been described. No enhancement in up to 40 - 50% cases.
- Enhancement is more common when cord enlargement is present (i.e. more frequent in the subacute than acute or chronic stages).
- Enhancing area is less extensive than T2 signal abnormality and resolves with time.
- ± meningeal enhancement.
- Cord atrophy in late stages.

Zaleswki., *et al*. found that spinal cord MRI was performed in 68.6% of admissions and external imaging was interpreted for the others. The injected T1 sequence was performed in 89.4% of cases. The diagnosis of idiopathic ATM was confirmed in 18.1% of cases and 69.9% were related to other myelopathies [3].

Biochemical and cyto-bacteriological analysis of the CSF is systematic (proteins, glucose, immunoglobins, toxoplasmosis, and syphilis germs). It will find, among other things, a pleocytosis, an increase in Immunoglobin G.

It will completed by a biochemical, hematologic, hormonal blood test and the search for antibodies (Vitamin B12, HIV, TSH, C Reactive Protein, syphilis TPHA-VRDL, anticorps anti NMO, aquaporin-4 AQP4, ASCA, BGSA) [9].

Other examinations may be carried out in the search for an etiology, in particular a thoracic-abdominal-pelvic scan, para-neoplastic tests, and an ophthalmological examination.

In the etiological diagnostic approach, the pathologies associated with ATM should be classified into three main groups:

- Infectious pathologies: Viruses (herpes virus, Enterovirus), bacteria (borreliosis, syphilis, tuberculosis), parasites (hydatidosis).
- Autoimmune pathologies (antibody research).
- Para neoplastic pathologies (search for anti-neuronal antibodies).

Many differential diagnoses can be listed (Table 2): Spinal cord infarct, multiple sclerosis, neuromyelitis Optica, spinal dural fistula, Syringohydromelia [4,10,11].

Differential Diagnoses	Clinical and radiological description
Spinal cord infarct	- Patients present more often with motor signs than sensory signs without inflammatory syndrome or fever. - Spinal cord infarct deficit occurs in hours rather than days. - Hyperintense signal on T2WI on both sides of the median fissure "Owl eyes" sign in the axial plane and extending over several segments "Pencil" sign in the sagittal plane, with or without enlargement of the spinal cord. - DWI shows restricted diffusion mostly in the anterior vertebral artery territory.
Multiple sclerosis	- Relapsing and remitting clinical course in contrast with the rapid onset of deficits in transverse myelitis. - Peripheral hyperintense signal on T2WI that doesn't exceed 2 vertebral segments in length. - Lesions are less than 1/2 of cord cross-sectional area in the axial plane. - Associated intracranial lesions.
Neuromyelitis Optica	Visual symptoms. - Optic nerve enhancement. - Long segment of cord enlargement. - Limited brain lesions.
Spinal dural fistula	Increased T2 cord signal, and prominent flow voids (dilated veins) on cord surface are in favor of spinal dural Fistula.
Syringohydromelia	Absence of sensory, motor, and autonomic dysfunction - A central cystic lesion - Absence of enhancement.

Table 2: *Differential diagnoses with clinical and MRI findings.*

Treatment consist in high-dose intravenous steroid pulse therapy and physical therapy. When ATM have secondary etiology, it must be treated also. ATM evolution is marked by a better prognosis on children than adults are. In general, 1/3 of cases recover completely, the second third may suffer from residual spasticity and urinary dysfunction. Complete finally persistent deficit may occur in the last 1/3 of patients. West., *et al*. found a low recovery rate of 33 - 38% with a mortality of 8.8% [4].

Depending on the severity, plasmapheresis has also been proposed as a treatment without any real proof of effectiveness. The diagnosis is sometimes late and leads to a modification of the treatment. Zaleswki., *et al.* in their study found a delay of 9 months (range 1 - 288) between the onset of symptoms and diagnosis. In 24% of cases, treatment was modified as a result of the final diagnosis [3].

Conclusion

ATM remains a rare neurological condition and diagnosis is based on clinical signs and spinal cord MRI data. An etiological assessment is systematically performed to confirm the diagnosis with a view to appropriate treatment. It involves analysis of the CSF and blood tests for infection, paraneoplasia, and immunology. The diagnosis must be precise to avoid the use of anti-inflammatory treatment in the case of non-inflammatory myelopathies.

Conflict of Interest

All authors have declared that no financial support was received from any organization for the submitted work.

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