

Transverse Myelitis in Turner Syndrome

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Background: Transverse Myelitis (TM) is a clinical syndrome in which an immune-mediated process causes neural injury to the spinal cord. The TM may be the main characteristic of Multiple Sclerosis (MS) and may be part of the initial picture of the disease. It is known that Turner's Syndrome is closely associated to the presence of autoantibodies and autoimmune diseases. We describe a case of a patient with Turner Syndrome (TS) who presented TM and can be an initial presentation of MS.

Objective and hypotheses: Clinical report

Method: Clinical report

Results: a 15-year old teenager, female, white, coming from Gravataí - Rio Grande do Sul, began with manifestations of tiredness and loss of strength in the lower limbs evolving rapidly with sensorial loss, tetraparesis and hemodynamic instability requiring intubation and vasoactive drugs. After two days was transferred to the pediatric intensive care unit (ICU). Patient diagnosed with hypothyroidism at 2 years and Turner Syndrome at age 11, had been in use vitamin D&calcium, conjugated estrogen, progestogen, levothyroxine, oxandrolone and growth hormone. On examination, the patient was in regular condition, weight 65kg, Glasgow 15, isochoric and reagents pupils, flaccid tetraplegia and areflexia. MRI showed extensive neuraxial sign of change in the central region of the cervical spine (hypointense on T1 and T2). Assessed by the pediatric neurology was diagnosed with Transverse Myelitis. She received 7 cycles of plasmapheresis with little clinical response, leaving the secondary MT print. Also received pulse therapy with methylprednisolone for 5 days and Rituximab with partial improvement of the strength members and weaning vasoactive drugs and mechanical ventilation. The patient was discharged from the ICU 2 months after admission, remaining tracheostomized and bedridden. One month later then she was discharged with prednisolone and azathioprine to continue home care.

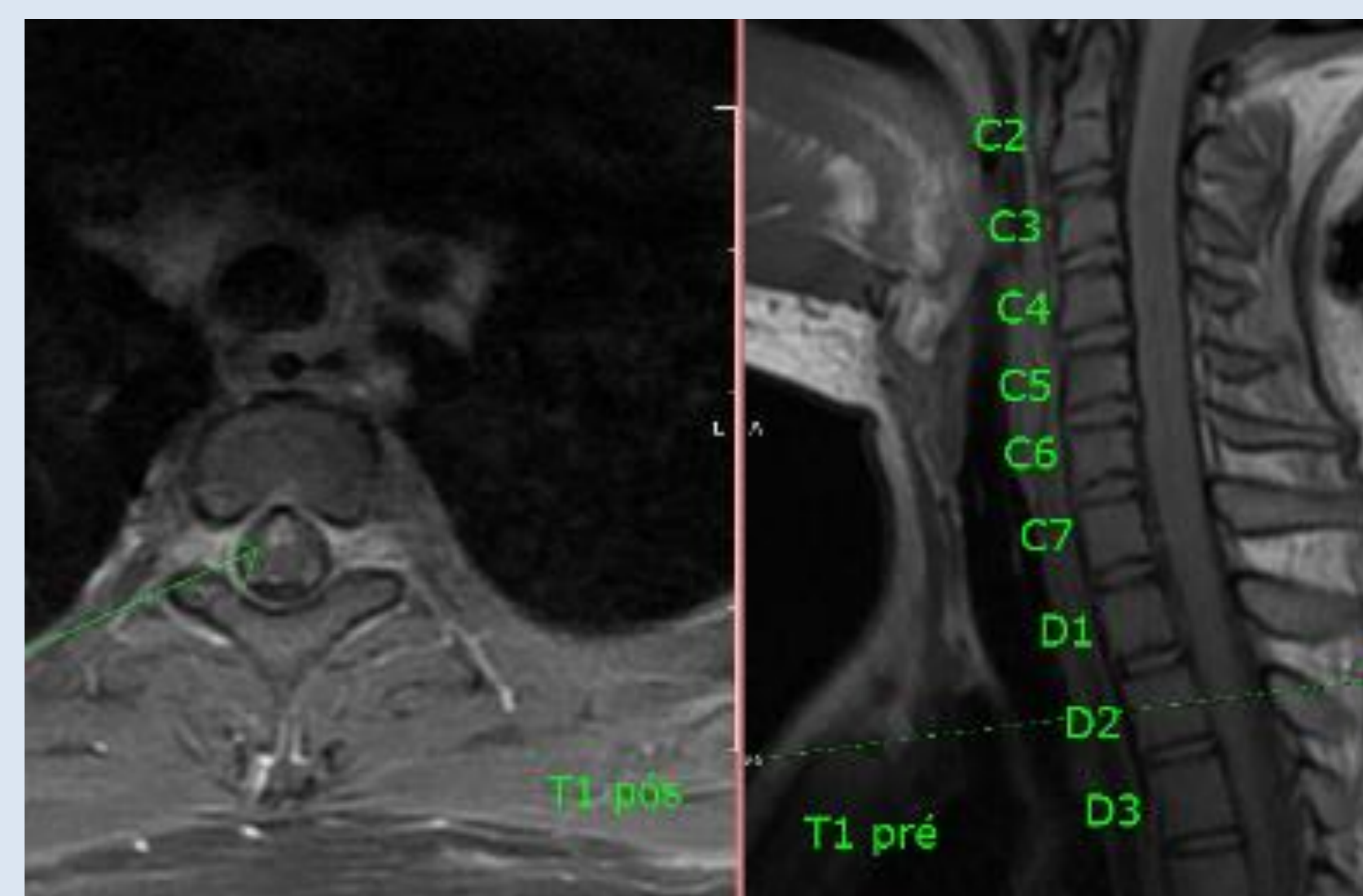


Figure 1



Figure 2

Figure 1 and 2. neuroaxial MRI

Conclusion: Turner's Syndrome is closely associated to the presence of autoantibodies and autoimmune diseases (AID), though its association with MS has been rarely reported. Despite the fact that the strong association between TS and AID is well known and has been widely studied, the underlying immunopathogenic mechanism remains partially unexplained. Recent studies have displayed how TS patients show an excess of immunogenic risk markers. This is evocative for a higher responsibility of X-chromosome abnormalities in the development of AID, and particularly of X-genes involved in immune response. Individuals with TS need life-long medical attention. Early diagnosis and regular screening for potential associated autoimmune conditions are essential in the medical follow-up of TS patients.

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