Cervical myelopathy and brachial plexopathy: an atypical presentation of Anti-Ma2 syndrome

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BACKGROUND

Paraneoplastic neurological syndromes (PNS) are a group of disorders caused by an abnormal immune response against tumor-expressed onconeural antigens, mimicking proteins of the central or peripheral nervous system. PNS clinical manifestations are quite heterogeneous. Encephalomyelitis, limbic encephalitis, and rapid progressive cerebellar syndromes are some of the presentations referred to as "high-risk phenotypes" [1]. These syndromes are associated with high risk antibodies (>70% associated with cancer), including Anti-Hu (ANNA-1), Anti-Yo (PCA-1), and Anti-Ma2 (anti-Ta), among the others. Anti-Ma2 antibody-associated PNS typically occurs in males, presenting with limbic, diencephalic, or brainstem dysfunction symptoms, while the spinal cord is not generally involved [2].

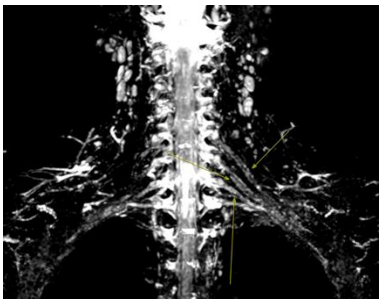
CASE REPORT

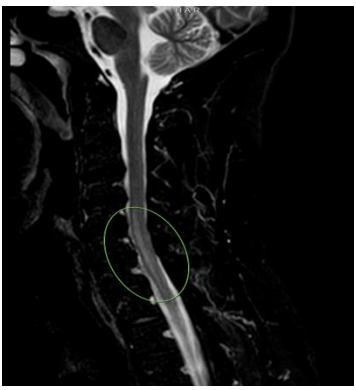
A 29-year-old male referred to our Neurology Unit for progressive bilateral upper limb weakness and fine motor dysfunction, accompanied by difficulty in swallowing and cervical pain. He had a history of callosal disgenesis with a mild intellectual disability and mixed testicular cancer, treated with orchiectomy and chemotherapy.

DIAGNOSTIC WORKOUT

Neurological examination

- Paraparetic gait
- Convergent strabismus of the right eye
- Spastic hypertonia of the lower limbs
- Weakness of the wrist and finger extensors, and of the thumb adductor muscles bilaterally
- Bimanual motor synkinesias ("mirror movements")
- Reduced deep tendon reflexes
- Bilateral Babinski and Hoffman signs





Electromyography

- Complete denervation of the left common extensor muscle of the fingers
- Neurogenic changes with chronic collateral reinnervation in the left radial flexor, right common extensor of the fingers, and bilateral first dorsal interosseous muscles

Laboratory tests

- Serum Anti-Ma2 antibodies +++

MRI of the cervical spine

- Necrotizing myelopathy at the C5-C7 level and brachial plexopathy

CONCLUSION

This case represents an unusual presentation of paraneoplastic syndrome associated with anti-Ma2 antibodies, since cervical myelopathy is not typically reported in this setting. In patients presenting with bilateral upper limb weakness with no clear evidence of central nervous system involvement, paraneoplastic syndrome should be carefully considered in the differential diagnosis, especially in the context of a history of testicular cancer. Early antibody testing, including Anti-Ma2, may facilitate timely diagnosis and appropriate management.

References
1. Graus F, Vogrig A, Muñiz-Castrillo S, et al. Updated Diagnostic Criteria for Paraneoplastic Neurologic Syndromes. Neurol Neuroimmunol Neuroinflamm. 2021 May 18;8(4): e1014.
2. Marsili L, Marcucci S, LaPorta J. Paraneoplastic Neurological Syndromes of the Central Nervous System: Pathophysiology, Diagnosis, and Treatment. Biomedicines. 2023 May 9;11(5):1406.