Combined central and peripheral demyelination mimicking Bing-Neel syndrome in Waldestrom macroglobulinemia: a case of successful treatment with Prednisone

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Case report: a 53 year-old woman was admitted to our hospital with a two weeks history of headache, muscle weakness of arms and legs, inability to walk and depression. Her medical history mentioned Waldestrom macroglobulinemia, chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), Aspergillus pneumonia and splenectomy. She was treated with Gabapentin, and she couldn't benefit from intravenous immunoglobulin (IVIg), because of a previous severe adverse reaction. Before hospital admission she walked single support, she had severe ataxia, bilateral foot dropping, hypoesthesia on extremities.
On admission her clinical examination revealed tetraparesis with absent reflexes, hypopallesthesia and sensory level at T12 without bladder dysfunction. She became bedridden and developed mild cognitive impairment.
Routine haematological, microbiological and virological investigations were negative, except for hyperproteinorachia. Magnetic Resonance Imaging (MRI) of the spinal cord showed an extensive myelopathy (7 cm from medulla oblongata to C5). On MRI of the brain cerebral white matter lesions hyperintense on T2 sequences with contrast enhancement on the right optic tract and parietal region, and bilateral temporal lobes (Fig.1, Fig.3 right). Electrodiagnostic studies indicated a severe symmetrical demyelinating neuropathy. Total body Computed Tomography (CT) was negative. Bing-Neel syndrome was proposed as a differential diagnosis, but it was excluded trough cytological cerebro-spinal fluid analysis. A diagnosis of CCPD was achieved and Prednisone 25 mg/die was administred.

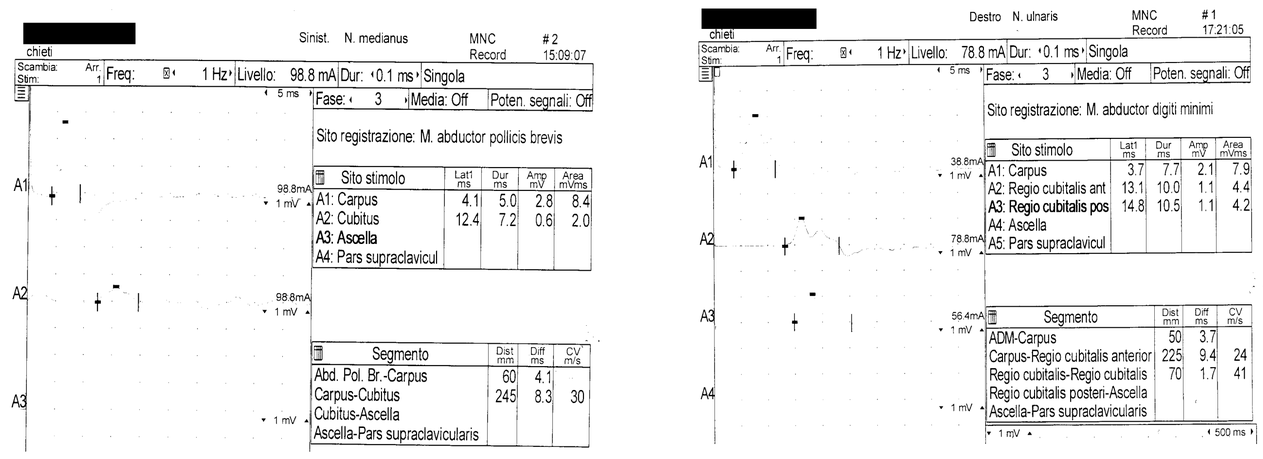

Fig.2. Electrodiagnostic studies showing CIDP features in the median and ulnar nerves.

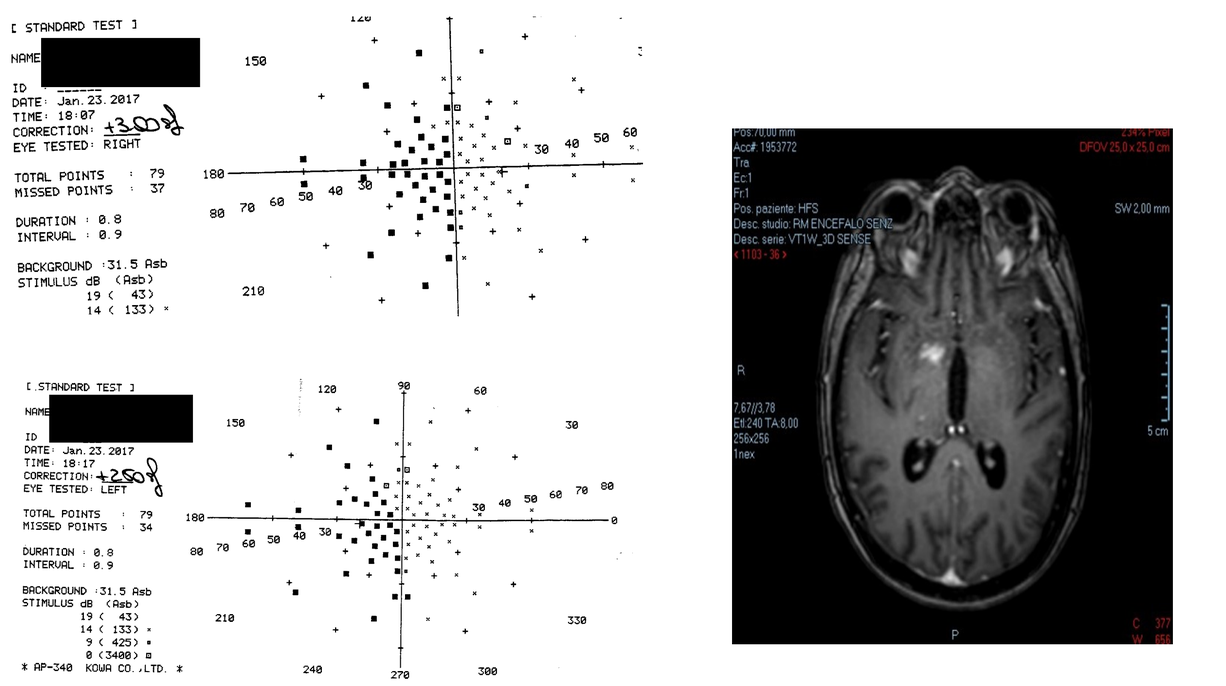

Fig.3. On the left campimetry showing hemianopsy; on the right hyperintensities of both optic nerves

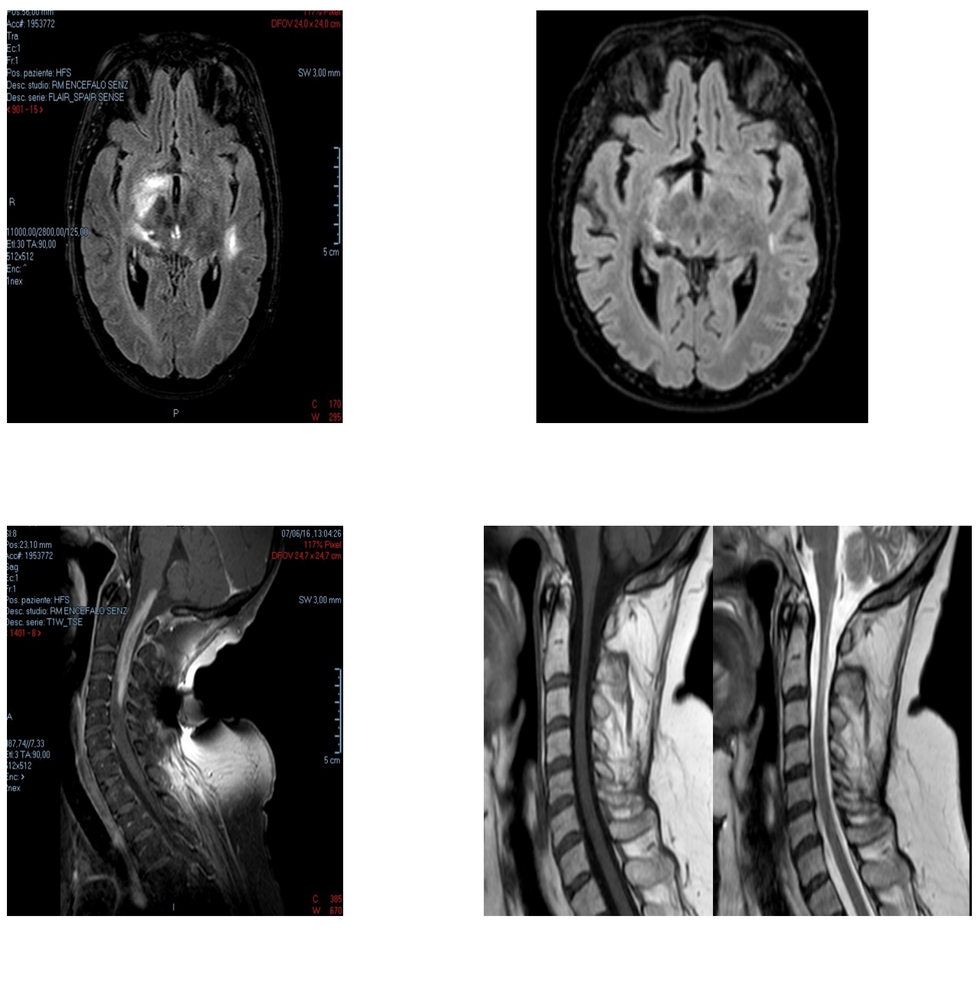

Fig.3. MRI changes before (on the left) and after (on the right) treatment with steroids.

Results
After a steroid treatment the patient could stand upright and walk with support; MRI of the brain and spinal cord showed dramatic regression of the T2 hyperintensities, with thinning of the spinal cord. Electrodiagnostic studies indicated improvement in compound motor action potential (CMAP) amplitude of the right ulnar nerve.

Discussion and conclusions: In accord with cases reported in literature, our patient presented clinical, laboratory and neuroimaging features that are atypical for multiple sclerosis with peripheral nerve demyelination which is indistinguishable from CIDP.
It has been reported that IVIg and plasma exchanges are beneficial for CCPD patients, while corticosteroids have a limited response. Although our patient couldn't beneficiate from IVIg, she dramatically improved with Prednisone therapy. We propose prednisone therapy as a valid choice in CCPD patients who cannot be treated with IVIg or plasma exchange; we suggest that immunomodulatory drugs might influence autoimmune response to antigens in the central and peripheral nervous system. Further studies are needed to confirm these observations.

References

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