case report

Paraneoplastic neuromyelitis optica spectrum disorder associated with stomach carcinoid tumor

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Neuromyelitis optica (NMO), or Devic's syndrome, is an autoimmune central nervous system demyelinating disorder primarily affecting the spinal cord and the optic nerves. It is characterized by the presence of NMO antibodies, alongside clinical and radiological findings. NMO and NMO-spectrum disorders (NMO-SD) have been reported in autoimmune disorders, and are infrequently described as a paraneoplastic syndrome with cancers of lung, breast, and carcinoid tumors of the thyroid. We report a patient who presented with severe vomiting, blurring of vision, vertigo, diplopia, left hemiparesis and hemisensory loss and ataxia. She was found to have a longitudinally-extensive demyelinating lesion extending from the medulla to the upper cervical spinal cord on MRI. Her gastric endoscopy revealed carcinoid tumor of the stomach, and classic paraneoplastic antibodies in the serum were negative. She had extremely high serum gastrin level and high titer of NMO IgG autoantibody. The patient made an excellent recovery with tumor resection and immunotherapy, with both clinical and radiological improvement. On rare instances, NMO or NMO-SD may present as a paraneoplastic neurological syndrome associated with carcinoid tumor of the stomach.

KEYWORDS: Demyelinating disease (CNS); Devic's syndrome; Autoimmune diseases; Paraneoplastic syndrome; Carcinoid tumor associated with paraneoplastic

Neuromyelitis optica (NMO), or Devic's disease, is an autoimmune, inflammatory central nervous system (CNS) syndrome, characterized by longitudinally extensive transverse myelitis, optic neuritis, and the presence of NMO-IgG autoantibodies.[1] The term NMO-spectrum disorder (NMO-SD) is used to include limited forms of NMO. (1) NMO could coexist with other autoimmune disorders,[2–4] and, rarely, may represent a paraneoplastic phenomenon.[5] We present a case of possible paraneoplastic NMO-SD in the setting of gastric carcinoid tumor. To our knowledge, the association of gastric carcinoid tumors with NMO or NMO-SD has not previously been reported.

CASE REPORT

A 38-year-old woman presented with severe vomiting, vertigo, blurred vision, diplopia, ataxic gait and left-sided numbness of two months' duration. Neurological examination demonstrated bilateral paleness of optic discs, horizontal nystagmus to left, left hemiparesis and hemisensory loss, left Babinski's sign, and gait ataxia.

MRI brain and spine revealed an enhancing hyperintense lesion extending from the medulla to the C2 cord segment (Figure 1A). Visual evoked potentials displayed bilaterally prolonged P-100. Routine blood investigations, ESR, autoimmune work up (ANA, Anti-DNA, SSA, SSB, ANCA, anti-phospholipid antibodies), viral serologies (HIV, HCV, HBV,

CMV, HSV 1 and 2), and VDRL were normal or negative. CSF analysis (cells, protein, sugar, AFB stain, bacterial cultures), and serum paraneoplastic antibodies (Hu, CV2, Ri, Ma2, amphiphysin), tested at Mayo Medical Laboratories, were negative. Serum B12 was low (83 pmol/L), and NMO-IgG was positive with high titer.

The patient had gastric endoscopy for evaluation of persistent vomiting, which revealed five gastric polyps at the antrum, treated with gastric ablation, and biopsy showed type I gastric carcinoid. Serum gastrin level was extremely high.

The patient received a five-day course of intravenous methylprednisolone (1gm/day), followed by

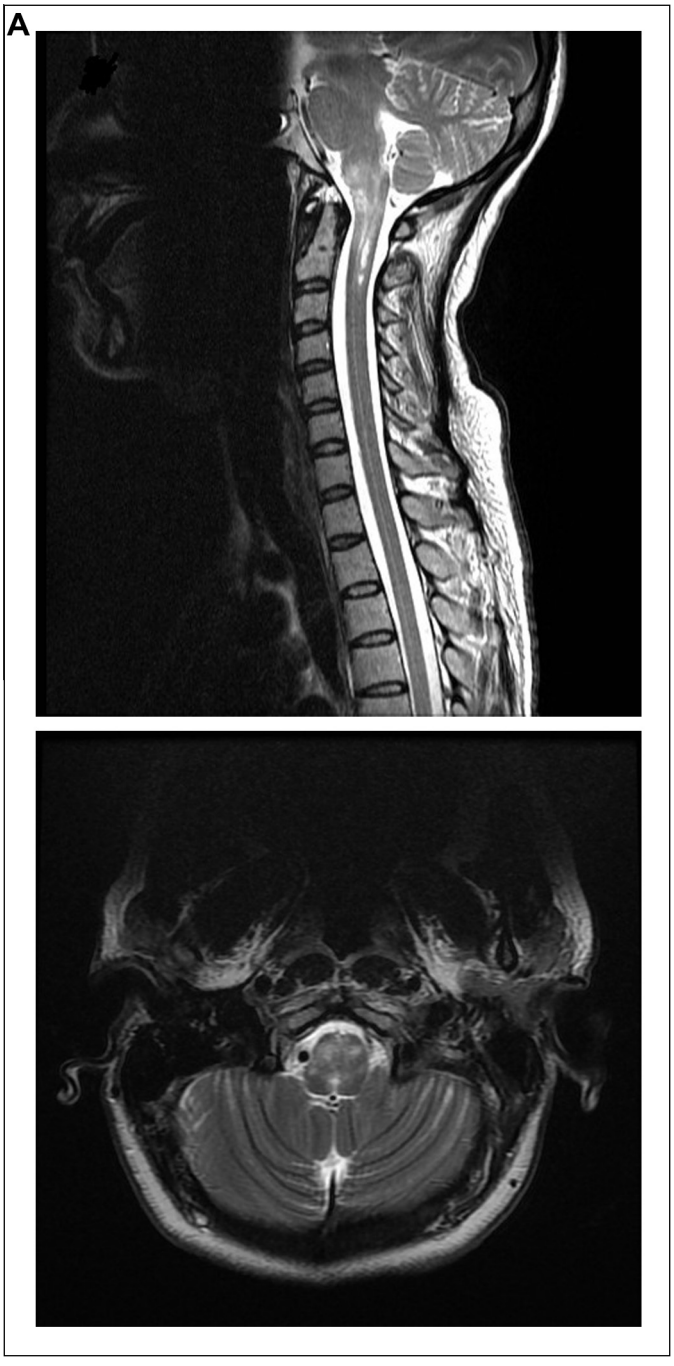

Figure 1A. Pre-treatment sagittal and axial T2-weighted MRI of medulla and cervical spine, showing diffuse, ill-defined hyperintense lesion involving the medulla oblongata and the upper cervical cord.