Neuromyelitis optica (NMO), or Devic’s syndrome, is an autoimmune, inflammatory central nervous system (CNS) syndrome, characterized by longitudinally extensive transverse myelitis, optic neuritis, and the presence of NMO-IgG autoantibodies. The term NMO-spectrum disorder (NMO-SD) is used to include limited forms of NMO. (1) NMO could coexist with other autoimmune disorders, and, rarely, may represent a paraneoplastic phenomenon. 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.
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
intravenous immunoglobulin (2 gm/kg over two days). This resulted in marked improvement in her neurological symptoms. She was discharged on prednisolone, azathioprine, and vitamin B12 replacement. Follow-up MRI showed significant regression of the previously observed lesion (Figure 1B), and several gastric endoscopies demonstrated no recurrence of the tumor. Serum gastrin returned to normal.

**DISCUSSION**

NMO is an autoimmune, inflammatory, demyelinating condition of the CNS, characterized by severe attacks of transverse myelitis and optic neuritis, with at least two of three supportive criteria, consisting of NMO-IgG seropositivity (directed against aquaporin-4 water channels), MRI spinal cord lesions extending more than three vertebral levels, and onset MRI brain

---

**Figure 1B.** Sagittal and axial T2-weighted MRI obtained after one year of therapy, showing resolving lesion at the cervicomedullary junction and upper cervical cord.
not meeting diagnostic criteria for multiple sclerosis. NMO-SD is a term also used to include limited forms of NMO, such as isolated single or recurrent forms of transverse myelitis, unilateral or bilateral optic neuritis, Asian optic-spinal multiple sclerosis, and NMO associated with systemic autoimmune disorders. These include systemic lupus erythematosus, Sjögren’s syndrome, and myasthenia gravis, and signify the coexistence of two autoimmune disorders rather than a vasculitic complication related to the systemic disease. Although rare, NMO may represent a paraneoplastic phenomenon associated with various cancers, including thymoma, breast, and thyroid tumors.

Carcinoids are neuroendocrine tumors that originate from the enterochromaffin cells of the gastrointestinal tract (74%), bronchial system (25%), and other less frequent organs. These tumors cause symptoms by local mass effect, fibrosis, metastasis, or secretion of bioactive substances, including various hormones, cytokines and growth factors. As a result, patients may present with a carcinoid syndrome, causing cutaneous flushing, diarrhea, abdominal pain, cardiopulmonary effects, and various endocrinological syndromes.

Carcinoid tumors may present with neurological symptoms, which, in the majority of cases, are due to endocrine changes, such as myopathy or metastases. Carcinoid-related paraneoplastic neurological syndromes have occasionally been described in isolated case reports. These include Lambert Eaton myasthenic syndrome, cerebellar degeneration, limbic encephalitis, sensory neuropathy, autonomic dysfunction, and myelopathy with brain stem encephalitis.

Our patient was diagnosed with NMO-SD, and was found to have gastric carcinoid tumor. While this occurrence could be incidental, it may represent a possible paraneoplastic effect of the cancer. She made an excellent recovery with treatment of the neurological condition, as well as tumor ablation, with no recurrence on prolonged follow up.

Pittock et al. reported several types of neoplasms in patients with NMO-SD and NMO IgG seropositivity, including breast carcinoma, thyroid Hürthle cell, carcinoid, B-cell lymphoma, and pituitary somatotropinoma. They propose that tumor cells may express onconeural antigens that can trigger an aquaporin-4 immune response. Indeed, aquaporin-4 is expressed in many tissues outside the CNS, such as skeletal muscle, lungs, and stomach parietal cells.

In addition, several cytokines, especially interleukin 6 (IL-6) are secreted directly by carcinoid tumors. By enhancing the production of various acute phase proteins such as serum amyloid, C-reactive protein, and fibrinogen, IL-6 has an important role in the development of inflammation. Similarly, NMO has been associated with a major activation of humoral immunity, and the CSF has shown significant increases in IL-5, IL-6, and IgM secreting cells.

CONFLICT OF INTEREST
None declared.

REFERENCES