ANTI-NMDAR ENCEPHALITIS ASSOCIATED WITH IMMATURE TERATOMA AND GLIOMATOSIS PERITONEI: A THERAPEUTIC CHALLENGE

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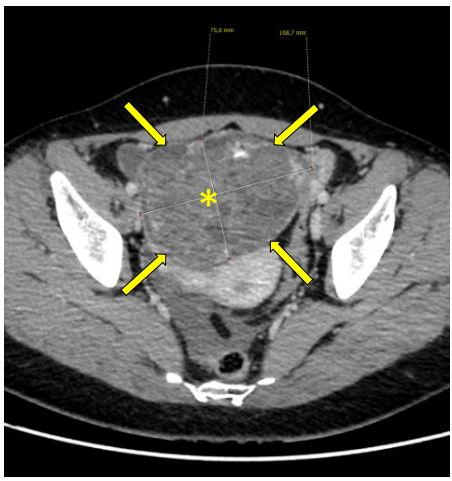
INTRODUCTION

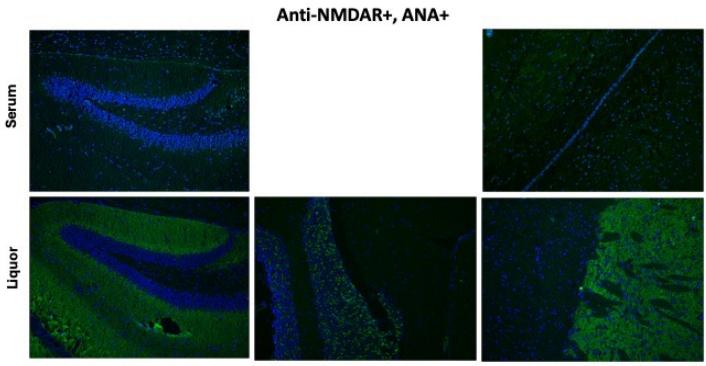
Anti-NMDAR encephalitis is a rare autoimmune encephalitis, with an incidence of 1 per million person-years. Tumors, mostly ovarian teratomas, are known triggers of NMDAR autoimmunity. In most cases, teratomas are mature; immature teratomas, which contain foci of immature neural tissue, have been described in 11.8% of cases. To our knowledge, the presence of gliomatosis peritonei in the context of anti-NMDARE is unprecedented and presents unique diagnostic and therapeutic challenges. Typically, treatment of anti-NMDARE involves immunotherapy and surgical removal of the underlying tumor. However, the optimal management of immature teratomas—particularly in cases complicated by gliomatosis peritonei—remains a subject of ongoing debate.

METHODS: Case Report.

RESULTS

This is the case of a 28-year-old female who underwent surgery to remove a pelvic mass, which was found to be a high-grade ovarian immature teratoma. One month later, the patient had two tonic-clonic seizures and was thereby hospitalized. The following days, she developed speech alterations evolving into mutism, cognitive impairment, abnormal behavior (agitation and disinhibition), movement abnormalities (oral and facial dyskinesias) and signs of dysautonomia. Brain MRI was normal. CSF analysis showed pleocytosis and high-titer positivity for NMDAR-antibody. The diagnosis of definite anti-NMDARE was made. The patient was initially treated with high-dose I.V. methylprednisolone for 5 days, then with I.V. immunoglobulin for 5 days and subsequently with two infusions of 1000 mg of Rituximab, with significant neurological improvement. Two diagnostic laparoscopies with multiple biopsies showed peritoneal deposits characterized by immature glial and neural tissue, related to the known immature teratoma. Chemotherapy with PEB regimen was therefore initiated. Her cognitive performances further improved.





«Fluorescence of the stratum moleculare of the hippocampus and the stratum granulosum of the cerebellum»

DISCUSSION

Management of anti-NMDAR encephalitis associated with immature ovarian teratoma require a multidisciplinary approach involving neurologists, gynecologists, and oncologists. In selected patients, in addition to immunotherapy and tumor removal, chemotherapy represents a valuable therapeutic option.

CONCLUSIONS

Our case report provides practical evidence regarding the therapeutic management of anti-NMDAR encephalitis associated with immature ovarian teratomas.

BIBLIOGRAPHY: Bost, C., Chanson, E., Picard, G. *et al.* Malignant tumors in autoimmune encephalitis with anti-NMDA receptor antibodies. *J Neurol* **265**, 2190–2200 (2018).

DISCLOSURES: The Authors have nothing to disclose.

24-28 Ottobre 2025 Padova Congress
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