Intrathecal NMDA receptor antibody reactivation due to new ovarian teratoma in patient with previously resected ovarian teratoma for anti-NMDAR encephalitis

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BACKGROUND

Anti-NMDAR encephalitis, an autoimmune encephalitis caused by CSF IgG antibodies against the GluN1 NMDA receptor (NMDAR) subunit, typically affects young adults and children, predominantly females harboring a mature teratoma. Antibody titers in CSF and serum are higher in patients with teratoma, and titer changes in CSF and serum are related to relapse. Relapses occur in 12-20% of cases. Patients either unresponsive to treatment or experiencing relapses should be reassessed for a contralateral or recurrent teratoma.

CASE REPORT

A 32-year-old female with history of clinically resolved NMDARE treated with left ovarian teratoma resection and methylprednisolone, IVIG, and rituximab presented with eclampsia 1.5 years after initial NDMARE diagnosis. The patient was unaware she was pregnant, and the infant eventually died of eclampsia after initial NDMARE diagnosis. The patient was treated with left ovarian teratoma resection and methylprednisolone due to clinical relapse.

OBJECTIVE

To describe the unusual presentation of a woman successfully treated for anti-NMDAR encephalitis (NMDARE) with ovarian teratoma resection and immunotherapy presenting with biochemical evidence of intrathalamic immune reactivation leading to the discovery of a new ovarian teratoma.

IMAGING

Relapse in encephalitis is defined as the new onset or worsening of symptoms occurring after at least two months of improvement. Relapses in NMDARE-may occur multiple times and have a higher frequency in patients without a tumor. Compared with the initial episode, relapses are less severe.

DISCUSSION

The presence of a teratoma not detected or untreated during the initial episode, or the possibility of tumor recurrence, should be considered in patients with a relapse. A relapse presents as the classic syndrome in only 31% of patients, the majority being incomplete or atypical syndromes.

Antibodies may persist for several years in the serum and CSF of asymptomatic patients after clinical recovery albeit at lower titers than during the initial episode. Self-contained meningeal germinal centers have been purported as a source of antibody synthesis without pathogenic effect on the brain. Relapse is associated with increase in CSF titers.

REFERENCES


CONCLUSION

Our case demonstrates an unusual presentation of immune reactivation in anti-NMDAR encephalitis without evidence of clinical relapse leading to the discovery of a new contralateral teratoma. The role of antibody and tumor surveillance in asymptomatic patients after recovery from NMDARE requires further study.

DISCLOSURES

The authors have no disclosures.