

Objective

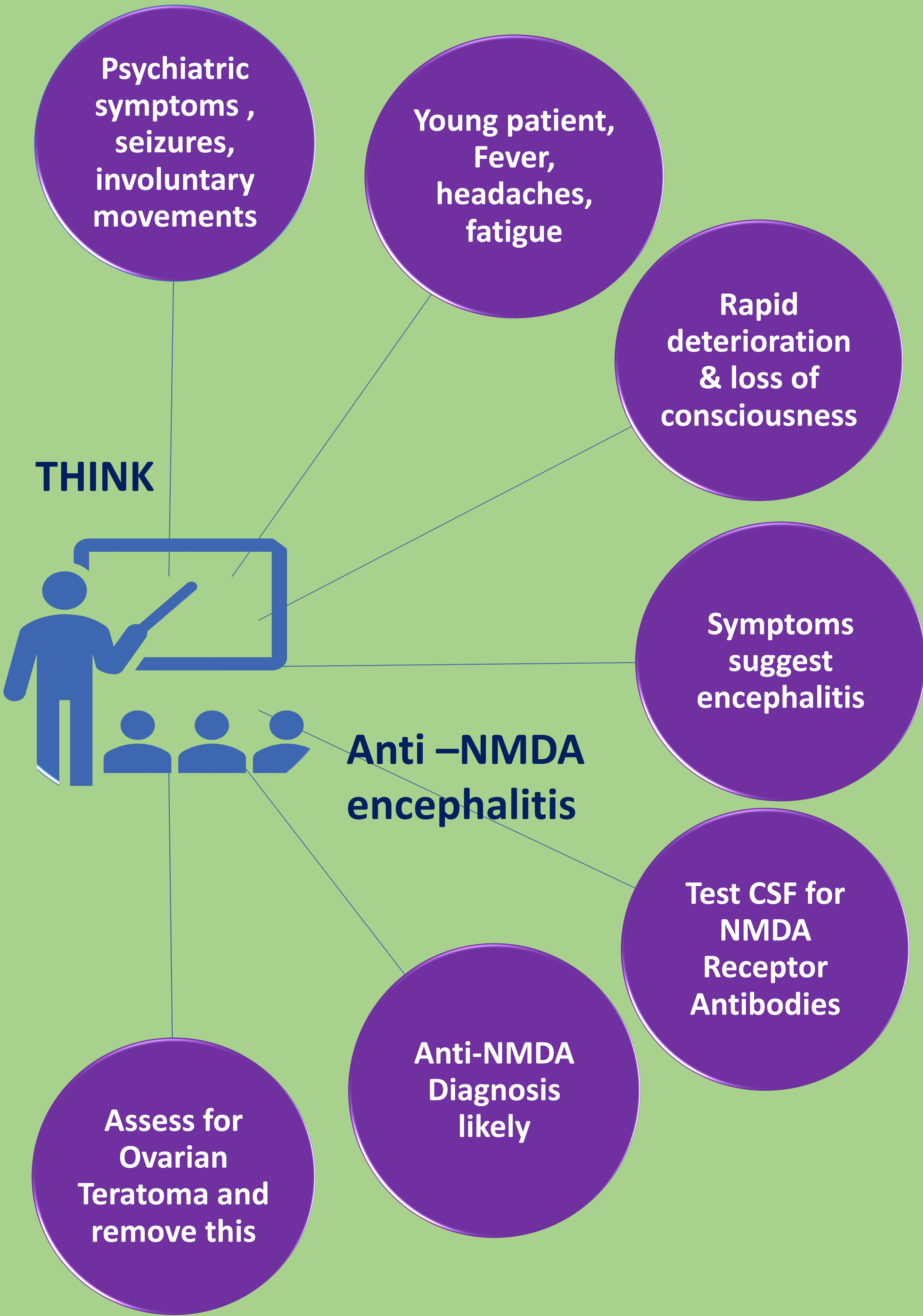
N-Methyl D-Aspartate (Anti NMDA) receptor encephalitis occurs in young women and is associated with ovarian teratoma. NMDA Encephalitis is an autoimmune disease with neurological and psychiatric symptoms. It is associated with significant morbidity.

It is a paraneoplastic syndrome associated with IgG antibodies affecting NDMA receptors in the brain. In cases of encephalitis in young women, NMDA receptor encephalitis secondary to an ovarian tumour should be considered as the diagnosis and discussed with a gynaecologist. It is likely that removal of the tumour is beneficial as this will eradicate the source of autoantibodies and aid recovery.

Case

A 16-year-old female presented to A&E following an unexplained collapse at home, where she became unconscious with irregular breathing. Prior to admission she had a headache for 2-3 days associated with dizziness. She was disoriented, had paranoid thoughts and repetitive speech. She was previously fit with no medical or mental health concerns. She was transferred to the intensive care unit (ICU) and had a normal CT head and inconclusive blood tests. EEG demonstrated encephalopathic changes with bilateral bursts of slow activity.

She was diagnosed with encephalitis and a CT scan suggested she may have a small ovarian teratoma. She was taken to theatre for a diagnostic laparoscopy and the ovary appeared normal. It was biopsied and histology was reported as inconclusive. She did not make any improvements in ICU and later had a PET scan; this again demonstrated a small ovarian teratoma. She had normal tumour markers, but NDMA receptor antibodies were positive. She was treated for NMDA receptor encephalitis and had a right oophorectomy. She was discharged 3 weeks later and histology confirmed a mature cystic ovarian teratoma.



Discussion

Anti NMDA receptor encephalitis is a potentially fatal condition associated with significant morbidity. Patients typically present with significant psychiatric symptoms as well as seizures and involuntary movements. Early features include fever, headache and fatigue. This rapidly progresses to unresponsiveness and can cause hypoventilation and dysautonomia. Anti-NMDA antibodies are present in cerebrospinal fluid (CSF) and this is an important clinical feature. Anti-NMDA receptor encephalitis usually occurs among young women and is often associated with ovarian tumours and more commonly mature teratomas.

- Early Tumour Removal

- Anti-NMDA Receptor encephalitis associated with ovarian teratoma is common.
 - Early tumour removal may improve prognosis
- Treatment

- Steroids
 - Plasma Exchange (plasmapheresis)
 - Intravenous immunoglobulin (IVIG)
- Potential Difficulties

- Rare presentation
 - Gynaecologists must have a central role in management
 - Teratomas can be microscopic & not detected in routine imagine

Conclusion

Anti NMDA receptor encephalitis is a very rare presentation associated with ovarian teratoma. It should be considered as a diagnosis in young women presenting with encephalitis. It is thought that removal of the teratoma improves recovery and long-term outcomes. Imaging can appear negative but a microscopic teratoma can still cause Anti-NMDA receptor encephalitis.

References

• Delangle R, Demeret S, Canlorbe G, Chelon L, Belghiti J, Gonthier C, Nikpayam M, Uzan C, Azaïs H. Anti-NMDA receptor encephalitis associated with ovarian tumor: the gynecologist point of view. Arch Gynecol Obstet. 2020 Aug;302(2):315-320. doi: 10.1007/s00404-020-05645-9. Epub 2020 Jun 17. PMID: 32556515.

• Yanai S, Hashiguchi Y, Kasai M, Fukuda T, Ichimura T, Yasui T, Sumi T. Early operative treatment of anti-N-methyl D-aspartate (anti-NMDA) receptor encephalitis in a patient with ovarian teratoma. Clin Exp Obstet Gynecol. 2015;42(6):819-21. PMID: 26753495.

• Abdul-Rahman ZM, Panegyres PK, Roeck M, Hawkins D, Bharath J, Grolman P, Neppe C, Palmer D. Anti-N-methyl-D-aspartate receptor encephalitis with an imaging-invisible ovarian teratoma: a case report. J Med Case Rep. 2016 Oct 24;10(1):296. doi: 10.1186/s13256-016-1067-4. PMID: 27776544; PMCID: PMC5078958.