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A Rare Case of Ovarian Teratoma - associated anti-NMDA Receptor Encephalitis

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Introduction

Anti-N-Methyl-D-aspartate receptor (anti-NMDAR) encephalitis associated with an ovarian teratoma is an under-recognised, rare and sinister immune-mediated paraneoplastic syndrome that predominantly affects young females. It was first described in the medical literature in 2005, and since then approximately 800 cases have been reported[1]. Due to the clinical presentation of neuropsychiatric symptoms, it is commonly misdiagnosed, delaying treatment and increasing the morbidity and mortality of this condition.

Case Report

Presentation

- 28 year old G0 presented to the Emergency Department with amnesia, confusion, and agitation. She was medically cleared and admitted to the psychiatric ward with dissociation secondary to an acute stress reaction.
- Patient's agitation increased, with periods of reduced consciousness prompting medical review. The patient's level of consciousness continued to deteriorate and she then became febrile. Bloods showed raised inflammatory markers. The patient was admitted to the Intensive Care Unit due to the low level of consciousness.
- A CT- Chest, abdomen and pelvis showed a left adnexal mass. A transabdominal pelvic ultrasound showed a complex cystic mass with solid components, measuring 69mm by 39mm. An MRI of the pelvis favoured the diagnosis of a mature cystic teratoma.
- Whilst undergoing these investigations, a positive result returned for anti-NMDA receptor antibodies.

Management

- Laparoscopic left Salpingo-oophorectomy was performed the next day. Histopathology confirmed mature cystic teratoma of the ovary.
- Five days of Intravenous Immune Globulin, with ongoing monthly infusions for a minimal of six months.
- Short course of plasmaphoresis, high dose Prednisolone.

Outcome

- The patient's recovery was complicated by a prolonged ICU admission with ventilatory support via tracheostomy
- Encephalitis causing autonomic dysregulation, rigidity and seizure activity
- Bowel ischemia requiring multiple laparotomies and small bowel resection
- Right iliac deep vein thrombosis requiring an inter-hospital transfer for placement of Inferior Vena Cava filter for three weeks and ongoing anticoagulation.
- Heparin-induced thrombocytopaenia and gastrointestinal bleeding secondary to the anticoagulation, resolved whilst an inpatient.
- She is currently neurologically intact and is undergoing rehabilitation.

Discussion

When ovarian teratoma's contain neural tissue, it triggers an immune response resulting in an overproduction of anti-NMDAR antibodies[1].

Clinical Presentation

Initial symptoms are viral-like with headaches, nausea, fever, vomiting and fatigue.
Neuropsychiatric symptoms then develop:

- early stage: confusion, memory loss, paranoia, hallucination, mood disturbance, seizures and movement disorders such as facial twitching. At this juncture, many patients are diagnosed with psychiatric disease.
- late stage: decreased responsiveness, hypoventilation and autonomic instability[2].

Patients can rapidly deteriorate neurologically, requiring ICU admission and ventilatory support[1].

Diagnosis

Evaluation of serum and CSF for anti-NMDAR antibodies and imaging, particularly transvaginal ultrasound, are essential components for diagnosis[1].

Management

Urgent removal of any ovarian mass is the priority, as well as immunotherapy via IVIg, corticosteroids and plasma exchange. This combined approach results in the most rapid recovery[3].

Problems arise when there is negative pelvic imaging, in the setting of NMDAR encephalitis. There are case reports of oophorectomy performed despite negative imaging that histology has then revealed an occult teratoma, and it's removal then providing a therapeutic response[4].

The systematic review by Acien et al[1] recommends a focused transvaginal ultrasound firstly. If this is inconclusive, proceed with full immunotherapy and assess the patients response before performing bilateral salpingo-oophorectomy[1].

References

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