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Acute encephalitis secondary to an ovarian teratoma

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Introduction

Anti-N-methyl-D-aspartate receptor (anti-NMDAR) encephalitis is a paraneoplastic syndrome associated with ovarian teratomas. It was discovered recently in 2007 and an increasing number of cases have been diagnosed, particularly in young women (Dalmau et al. 2011). Patients typically present with prominent psychiatric symptoms, seizures, involuntary movements and rapidly progress into unresponsiveness with central hypoventilation and dysautonomia (Iizuka et al. 2008). We present a rare case and a review of current management.

Case report

A 31-year-old woman presented to the neurologists with acute agitation and confusion. She had no past medical history and was not taking any regular medication. There was no history of substance abuse. Examination itself was unremarkable. On day 3, the patient became unconsciousness (GCS = 3) and was sedated and ventilated in Intensive Care. She also developed seizures, marked orofacial dyskinesia and involuntary movements of the upper neck, head and upper limbs.

Analysis of CSF was normal. MRI of the brain showed bilateral asymmetric swelling of the temporal lobe; EEG showed periodic discharges consistent with encephalitis. Other routine investigations to establish a cause for encephalitis were negative (Table I). Therefore a literature search was made, after which anti-NMDAR encephalitis was suspected (Vitalini et al. 2005; Dalmau et al. 2007; Sansing et al. 2007). A CSF and serum sample were sent to a US laboratory, where this antibody assay was available. Both tested positive for NMDAR antibodies. The gynaecologists were involved at this point and an MRI pelvis detected a 3×3 cm right ovarian dermoid cyst. Therefore, the patient was diagnosed with anti-NMDAR encephalitis associated with ovarian teratoma.

She was given high dose corticosteroids and plasma exchanges were done. On day 56, the patient had an uncomplicated unilateral right oophorectomy. Surgical histology confirmed the presence of neural tissue within a mature teratoma, with evidence of gliosis and an inflammatory infiltrate supporting a central nervous system directed immune response.

The patient made a dramatic recovery within 2 weeks of surgery. She regained consciousness and was vocalising, eating and able to walk with a frame. A month after intensive rehabilitation, the patient was discharged home, capable of independent daily living.

Discussion

Patients with anti-NMDAR encephalitis will most likely present to members of the neurology or psychiatry team before being referred to gynaecologists. Owing to its recent discovery, there may be a delay in diagnosis due to lack of familiarity. Furthermore, anti-NMDAR is not included with routine paraneoplastic antibody screen. Current literature suggests that a search for an ovarian teratoma should be undertaken following a positive antibody test. In the largest case series analysis, 41% of patients have no identifiable tumour (Dalmau et al. 2008). However, it has been recommended that these patients be screened for ovarian tumours 2 years following diagnosis (Dalmau et al. 2011).

Although no standard of treatment exists, it principally involves immunosuppression (steroids, chemotherapy, monoclonal antibodies, plasma exchanges) and removal of any existing ovarian tumour (Iizuka & Sakai 2008). Oophorectomy was chosen over a cystectomy to ensure that no tumour remnant could persist as an antigen source. Patients who received treatment within 4 months of initial presentation tended to have better outcomes (full recovery, residual mild neurological deficits) compared with those having delayed treatment ($p = 0.004$). It was also shown that this group of patients recovered more quickly (8 weeks vs 10–11 weeks) (Dalmau et al. 2008; Katakoa et al. 2008; Seki et al. 2008).

Conclusion

Early recognition of this rare disorder can be encouraged by increasing awareness among relevant disciplines. The diagnosis of anti-NMDAR encephalitis should prompt a search for an ovarian teratoma. Advice should also be sought from an interdisciplinary team with experience of managing these rare and complex cases.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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Table I. Summary of investigations.

Investigations		Results
Lumbar puncture	CSF analysis	Normal
Radiological imaging	MRI brain	Bilateral asymmetric temporal lobe swelling
	US/MRI scan (pelvis)	3×3 cm right ovarian dermoid cyst
	Whole body PET scan	Negative
	Whole body CT scan	Negative
Tumour markers	CA125, β-HCG, AFP, LDH, CEA	Normal ranges
Paraneoplastic antibodies	Anti-Hu, Anti-Ri, Anti-Yo	Negative
	Anti NMDAR	Positive
EEG	–	Periodic discharges consistent with encephalitis. Not status epilepticus
Immunological	Coeliac, ANA, ANCA, anti-smooth-muscle, anti-LKM, anti-mitochondrial, anti-cardiolipin, anti-VGKC, anti-TPO	Negative
Virological/bacteriological	VZV, HSV, HIV	Negative
	Whipple's, gonorrhoea, ASOT	Negative

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