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Case Report

Autoimmune Encephalitis with Ovarian Teratoma: A Case Presentation and Review

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Abstract

Introduction: Anti NDMAR encephalitis is relatively common autoimmune encephalitis characterized by complex neuropsychiatric feature and the presence of immunoglobulin G {IgG} antibodies against the NR1 subunit of the NDMA receptors in the central nervous system.

Case Report: This is a case of a 53-year-old woman who had small ovarian teratoma. She presented to the hospital with complaint of right lower abdominal pain. Ultrasound examination showed presence of dermoid cyst measuring 2.5 cm by 2.5 cm in the right ovary and no evidence of torsion.

She presented the second time with severe manifestations including fever, focal seizure, incoherent speech followed by onset of generalized tonic-clonic convulsion lasting for five minutes which progressed to status epilepticus. She was then sedated and intubated. Her medical history included hypertension and hypercholesterolemia and no history of epilepsy.

Investigations like full blood counts (FBC), white cell count (WCC) and inflammatory markers were unremarkable. Lumbar puncture and MRI brain showed autoimmune encephalitis whereas CT head scan did not show signs of intracranial bleedings. The presence of NMDAR antibodies was confirmed by serum testing.

Autoimmune encephalitis was diagnosed by NMDAR antibody triggered by an ovarian teratoma. Surgical intervention was decided upon by a multidisciplinary team involving gynecology, MICU, and neurology. 3 cm x 3 cm right ovarian dermoid cyst found during laparoscopy and the patient underwent laparoscopic bilateral salpingo-oophorectomy.

Postoperatively, the patient was continuously convulsing for which she was sedated, but gradually improved but frequent facial and right-hand twitch remain.

MRI repeated 2 months after surgery show no residual tumor.

She was discharged to rehabilitation with poor Glasgow coma scale of 3, tracheostomized on naso gastric tube for feeding.

Conclusion: The association of ovarian teratoma anti-NDMAR encephalitis is a serious and potentially fatal pathology. Heightened recognition of cognitive and behavioural change, diagnosis through TVU and subsequent tumor removal in addition to diagnostic confirmation through the presence of anti-NDMAR antibodies must be emphasized.

Keywords: *Autoimmune Encephalitis; Ovarian Teratoma; Anti-NDMAR Antibodies*

Abbreviations

MDT: Multi Disciplinary Team; Anti NDMAR: Anti-N-Methyl-D-Aspartate Receptor; MICU: Medical Intensive Care Unit

Introduction

Anti NDMAR encephalitis is relatively common autoimmune encephalitis characterized by complex neuropsychiatric feature and the presence of immunoglobulin G {IgG} antibodies against the NR1 subunit of the NDMA receptors in the central nervous system.

Case Presentation

This is a case of a 53-year-old woman who had had a small ovarian teratoma since 2019. On June 14th, she was presented to hospital with complaint of right lower abdominal pain. Ultrasound examination showed presence of dermoid cyst measuring 2.5 cm by 2.5 cm located in the right ovary and no evidence of torsion.

She came back on June 23rd with severe manifestations including fever, focal seizure, incoherent speech followed by onset of generalized tonic-o clonic convulsion lasting for five minutes which progressed to status epilepticus. She was then sedated and intubated. Her medical history included hypertension and hypercholesterolemia no previous history of epilepsy, while surgeries past have been done that include hysteroscopy as well as polypectomy.

Investigations like full blood counts (FBC), white cell count (WCC) and inflammatory markers were unremarkable. Lumbar puncture and MRI brain showed autoimmune encephalitis whereas CT head scan did not show signs of intracranial bleedings or haemorrhage. The presence of NMDAR antibodies was confirmed by serum testing.

Autoimmune encephalitis was diagnosed by NMDAR antibody triggered by an ovarian teratoma. Surgical intervention was decided upon by a multidisciplinary team involving gynecology, MICU, and neurology a 3x3 cm right ovarian dermoid cyst, a normal-looking right tube, left tube, and left ovary were found during laparoscopy, bilateral salpingo-oophorectomy were performed.

Postoperatively, she stayed 6 months in the hospital discharged on poor Glasgow coma scale score 3, tracheostomized and on nasogastric tube for feeding.

Discussion

Ovarian teratoma (OT) represents 20% of all ovarian neoplasms and are a common cause of paraneoplastic syndromes (PNS). Various tissues including teeth, hair, and neural tissue can be found in these tumors leading to different clinical presentations. OT is known to be linked with anti-NMDA-R encephalitis which is a well-known PNS. The last ten years have seen the detection of many new antibodies possibly associated with this disease thus revealing its immune-mediated nature [1].

Frequency and classification of ovarian teratoma-related paraneoplastic neurological syndromes

The frequency of OT-associated PNS has not been established well. In one Japanese single-center retrospective study among 343 patients, anti-NMDAR encephalitis was found in 1.17% cases of ovarian teratoma; while another study from Israel reported an incidence rate of 0.85%, among 233 patients over a period of 12 years respectively.

However, none did a complete analysis for other PNS except for anti-NMDAR encephalitis or elaborate on antibody screening [2].

Ovarian teratomas are classified into mature and immature types; the mature teratoma being more common and usually benign. Immature teratomas are less common but they may be malignant. This is important particularly in these tumors that have neural tissue because it may generate pathogenic antibodies thereby making them significant to PNSs [3].

Neurological syndromes of OT-related PNSs

Autoimmune encephalitis is the most common neurological syndrome related to OT, particularly anti-NMDAR encephalitis. Patients typically complain of a wide range of neurological and neuropsychiatric symptoms, including seizures, abnormal movements, and cognitive impairment. Less frequently, some gynaecological symptoms like lower abdominal pain are seen before the neurological symptoms. OT, therefore, is confirmed by further examinations of such symptoms [4].

Pathogenesis

Anti-NMDAR encephalitis is thought to involve antibody-mediated damage to the neurons. The antibodies formed against NMDARs on the teratoma cross the blood-brain barrier and attack the neurons. It has been estimated that roughly 37.4% of all anti- NMDAR encephalitis cases have OTs, in young women between the ages of 18 and 35 years. It appears that the interaction of immune and neural tissue within the teratoma is essential for the development of the encephalitis [4].

Diagnosis

A team of experts recommended criteria for the diagnosis of definite anti-NMDAR encephalitis, which could be made if the following were satisfied: rapid onset of at least four of six major symptom groups within less than 3 months; the major symptom groups include psychiatric symptoms or cognitive impairment, seizures, speech dysfunction, abnormal movements, decreased consciousness, autonomic dysfunction, or central hypoventilation; positive CSF IgG anti-GluN1 antibodies; and reasonable exclusion of other disorders. In most cases, anti-NMDAR encephalitis with OT presents severely [5].

Treatments and outcomes

Treatment for OT-related PNSs incorporates tumor resection, immunotherapy, and symptom control. Tumor resection was helpful for neurological progression-halting more than immunotherapy; some patients did not need additional immunotherapy at all.

Systemic and neurological complications must not pose a contraindication to surgery. Delayed resection of teratomas may foster the incessant presentation of antigens, thereby inducing long-lived plasma cells and increasing the affinity of antibodies. Such an approach would render late tumor resection and immunotherapy ineffective.

Prophylactic oophorectomy is not indicated in cases of anti-NMDAR encephalitis without detectable OT but can be considered in some patients who have a severe neurological involvement refractory to immunotherapy for more than 2 - 3 months [6,7].

Prognosis

The response to therapy and prognosis depend on the underlying immune mechanism. Patients with antibodies against intracellular (onconeural) antigens tend to have poorer responses than those with antibodies against surface or synaptic antigens. OT- associated anti-NMDAR encephalitis generally has a good prognosis, with partial or full recovery in about 80% of patients, though some cases result in severe disability or death. Most patients with PNSs experience full or near-full recovery after tumor removal and immunotherapy [8]. Long-term follow-up is essential to manage the potential relapses and neurological sequelae. Rehabilitative and psychological support may also be needed to cope with residual neuropsychological or psychiatric sequelae [9-11].

Conclusion

This complex, serious, and rare neurological disorder calls for collaborative efforts from different fields of medicine in its proper diagnosis and treatment. Further research and heightening of clinical awareness are required in order to increase the chances of a better prognosis for affected patients, as neurological complications can be so serious.

Conflict of Interest

I have no conflict of interest.

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