Ovarian teratoma-associated anti-NMDAR encephalitis in a 12-year-old girl

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SUMMARY

The association of ovarian teratoma and anti-N-Methyl-D-aspartate receptor (anti-NMDAR) is one of the most common autoimmune encephalitis syndromes and it is a serious and potentially fatal pathology that occurs in young women. This case report describes of a pediatric patient with anti-NMDAR encephalitis. A-12-year-old girl presented with abnormal behavior for one week came to Emergency Department of Sarawak General Hospital, Malaysia. She had psychotic spectrum symptoms including suicidal tendency. She was diagnosed with anti-NMDAR encephalitis as positive antibody was seen in her cerebrospinal fluid. She was treated with Injection Immunoglobulin. She turned out to have teratoma which was successfully removed later. Her progress was remarkable after the surgery with the Immunoglobulin. A multi-disciplinary team involving a psychiatrist, neurologist and gynaecologist liaised with intensivist to successfully manage the case and achieve the good outcome.

INTRODUCTION

The association between ovarian teratoma and anti-N-Methyl-D-aspartate receptor (Anti-NMDAR) is now recognized by most of clinicians. In a cohort study of Titulaer MJ et al.[4] with 577 anti-NMDAR encephalitis patients had observed that Asian and African-American patients found out that large group of the patients 220 (38%) had teratoma, especially in women 213(46%). Anti-NMDAR cases were rarely seen in girls of age younger than 12 years is only 4 (6%) and male patients 7(6%) in that study. This case report a serious and potentially fatal pathology named autoimmune encephalitis syndrome in a 12-year-old previously healthy girl. In this case, we successfully managed the anti-NMDAR encephalitis with previous unknown history of teratoma in a young girl by a competent multi-disciplinary management in order to share this uncommon pathology with other clinicians.

CASE REPORT

A 12-year-old girl who presented with a sudden development of abnormal behavior for one week came to the Emergency Department of Sarawak General Hospital. She was treated for some psychiatric problems in a private hospital last a few days ago. She had no medical illness before this incidence with no family history of psychiatric illness. History of illicit drug use and travel history were ruled out.

On examination, the patient was afebrile and her vital signs were normal and stable. Her Glasgow Coma Score was 13/15 on that time. All systemic examinations were unremarkable. All routine necessary blood tests were done and results were within normal limit. She was seen by psychiatric team to rule out psychosis and other psychiatric disorders while waiting for advanced investigations. Computerized tomographic brain scan was done and the result revealed there was no gross abnormality. Magnetic Resonance Imaging reported absence of space occupying lesion or any demyelinating lesions were detected. However, Electroencephalogram (EEG) result indicated an abnormal EEG with continuous delta wave suggestive of severe encephalopathy. She developed nystagmus movement for 10-20 seconds which was suspected to be a seizure while in the emergency department and she was given intravenous phenytoin to control her seizure. Her condition worsened as she lost consciousness after admission. We proceeded to lumbar puncture (LP) the next day. There have differential diagnoses like schizophrenia, substance abuse or malingering, the vigilant expertise of our neurological team, they looked for anti-NMDAR antibodies in the cerebrospinal fluid of the patient. LP result confirmed the presence of anti-NMDAR antibodies (positive) with plasma glucose of 0.6 and 1.4 in lymphocytes count. Again, the neurologist took care of the patient referred her to the Obstetrics and Gynaecology team in order to rule out ovarian tumour especially teratoma. Transabdominal ultrasound was done and there was a well-defined solid cystic lesion 5.5x5x4.7cm at vesico-uterine pouch with hyperechoic nodule and calcification. Tumour marker was taken and all were within normal limits.

Laparotomy with left ovarian cystectomy was performed and the sample was sent for histopathological examination (HPE), together with minimal peritoneal fluid for cytological examination. Both her fallopian tubes, the right ovary and uterus were normal. She was treated with intravenous (IV) immunoglobulin and IV methylprednisolone for 5 days followed by oral prednisolone tablets. Fig (1) showed HPE microscopic appearance. The microscopic feature shows that cyst wall is partly lined by keratinized stratified squamous (f)

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