Anti-NMDAR encephalitis in a 19 year old female patient with ovarian teratoma: A case report

Johnny El Hanna*, Codjo Quenum, Abdelilah Arsalane

Department of Obstetrics and Gynecology, Grand Hôpital de l’Est Francilien, France

ARTICLE INFO

Article history:
Received 20 March 2021
Accepted 7 June 2021
Available online 12 June 2021

Keywords:
Encephalitis
Autoimmune
Anti-N-Methyl-D-Aspartate
Ovarian teratoma

ABSTRACT

Background: Anti-N-Methyl-D-Aspartate encephalitis is a subcategory of auto-immune encephalitis. It is known for its aggressive presenting symptoms and rapid deterioration, yet it is treatment responsive. It is associated in 50% to ovarian teratoma.

Case: We report the case of a 19 year old female patient presenting for a psychiatric disorder of sudden onset with rapid deterioration. Neurologic imaging was in favor of encephalitis, and CSF studies revealed Anti NMDA receptors. Further abdominal imaging showed a right ovarian teratoma of 4 cm.

Conclusion: Anti-NMDA receptor encephalitis with ovarian teratoma is a rare entity with rapid deterioration. Early diagnosis, surgical resection and proper medical treatment are essential for the management of this disease.

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Introduction

Encephalitis is defined as brain inflammation with neurologic symptoms due to inflammation of brain parenchyma. It arises from either an infectious etiology or an autoimmune disorder [1]. The affected patients suffer from psychiatric symptoms with decreased level of consciousness, seizures and autonomic dysfunction, often requiring ventilatory support [2].

Vitalini et. Al first described in 2005 a case series of 4 patients with a pattern of paraneoplastic encephalitis and ovarian teratoma [3]. Later analysis by Dalmau et. Al suggested a model of autoimmne encephalitis associated with antibodies to NR2B and NR2A heteromers of the N-Methyl-D-Aspartate receptor (NMDAR) [2]. Case reports progressively increased as of 2007, showing association between auto-immune encephalitis and ovarian teratomas [4].

We present in this article the case of a 19 year old female patient suffering from anti-NMDAR encephalitis associated with a 4 cm ovarian teratoma.

Case presentation

This is the case of a 19 year old French patient of African descent, brought to the ER by her parents for minor headache of 2 days duration with low grade fever. Lab tests were done showing normal WBCs and CRP of 9. She was sent home with pain killers. She was brought back to the ER two days later, with intense headache, insomnia, dysartria and aggressive behavior. Panels showed no major modifications. Neurology department was consulted, they advised to hospitalize the patient and proceed to an urgent brain MRI.

Brain MRI showed no signal abnormalities on DWI and ADC sequences. No microbleeds or calcifications were seen on T2*. On T2 FLAIR 3D sequence, a small hyperintense lesion was seen over the right medial temporal area, on the coronal view. No abnormal contrast enhancing lesions on T1W images. EEG was not conclusive with diffuse microvoltage activity and no reaction to photic stimulation. EEG repeated two days later showed an epileptic activity with Theta rhythmic waves over the left hemisphere, lasting twenty seconds, followed by flat recording in post ictal phase. CSF analysis showed lymphocytic pleocytosis with normal protein, glucose and gram stain, and negative bacterial and viral PCR panels. Positive oligoclonal bands were seen in CSF as well in serum, with normal IgG index.

Autoimmune encephalitis was suspected. CSF antibody studies showed positive antibodies to the NMDA receptor. Patient was started on antiepileptic drugs, Solumedrol 1 g/day IV for 5 days

* Corresponding author.
E-mail address: jhanny.hanna@live.com (J. El Hanna).
dermatitis, endoderm and mesoderm origins. Less than 3% of
dermatitis are malignant. Management mainly includes
laparoscopic resection and extraction within a bag [7,8].
Diagnosis of encephalitis and searching for the etiology remains
complex, and more than 50% of encephalitis remain of unknown
etiology [5]. Diagnostic testing include lumbar puncture, brain MRI
and EEG. Diagnostic criteria include one major criterion: patient
presenting to medical attention with altered mental status lasting
≥24 h with no alternative cause identified, with >2 minor criteria:
Documented fever >38 degrees C within the 72 h before or
after presentation, generalized or partial seizures not fully
attributable to a preexisting seizure disorder, new onset of focal
neurologic finding, CSF WBC count ≥5/cubic mm^3, abnormality of
brain parenchyma on neuroimaging suggestive of encephalitis that
is either new from prior studies or appears acute in onset,
abnormality on electroencephalography that is consistent with
encephalitis and not attributable to another cause [1]. In the case of
our patient, symptoms upon presentation, brain MRI, EEG and
lumbar puncture were in favor of autoimmune encephalitis.

After the work of Vitaliani et al [3] and Dalmau et al [2]
between 2005 and 2007, a new category of paraneoplastic
encephalitis have emerged, associating severe encephalitis with
ovarian teratoma in young female patients and anti NMDA receptor
antibodies. Once anti-NMDAR encephalitis is suspected, an
abdominal CT scan is recommended, searching for an ovarian
teratoma cyst present in 50% of patients [4,6]. Treatment includes
surgical ovarian cyst resection. Other therapies include immuno-
therapy like corticosteroids, intravenous immunoglobulin, or
plasma exchange [9]. Our patient was put on anticonvulsants
after her first seizure (Keppra 500 mg 2%/day, Valium 10 mg every 6 h). Once the
MRI, EEG and CSF analysis were in favor of autoimmune
encephalitis, she was started on Solumedrol 1 mg/kg IV. The
detection of anti-NMDA receptor antibodies was an argument to
introduce IVIG (CLAIRIG) 2 g/kg given over 6 days. IVIG was
given over 2 cures before surgery, and 1 cure after surgery. Corticoste-
roids were maintained after the laparoscopic resection.

According to a systematic review in 2014 [4], improvement
with hospital discharge was acceptable after a median of 3 months.
Full recovery was noticed in 80% of patient. Our patient showed
minor improvement after 2.5 months, with hospital discharge after
3.5 months. Full recovery was achieved after 7 months.

Conclusion
Anti-NMDA encephalitis associated to ovarian teratoma is a rare
entity that carries high mortality and morbidity. The rarity of this
category often delays diagnosis, with a mean time to surgery
around 74 days in the case of mature teratoma [4]. This pathology
needs better understanding and more acknowledgment by
neurologists and gynecologists, in order to set early diagnosis
and proper management.

Declaration of Competing Interest
The authors report no declarations of interest.

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