Small Ovarian Teratoma Causes Anti-N-methyl-D-aspartate Encephalitis

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This is the case of a 22-year-old female who arrived at our institution after experiencing refractory insomnia, disorganized behavior, inappropriate laughter, and anorexia. Upon admission, a physical examination revealed mutism, irritability, and visual hallucinations. Infectious, metabolic, and other, alternative, causes for the presenting symptoms were excluded. Brain magnetic resonance imaging and chest and abdominopelvic computed tomography scan results showed no evidence of pathology. Due to there being a high clinical suspicion of paraneoplastic encephalitis, treatment was initiated with intravenous (IV) high-dose steroids and IV immunoglobulins. An endovaginal ultrasound was performed, which revealed a small atypical intraovarian dermoid cyst. The patient's laboratory tests were positive for anti-N-methyl-D-aspartate antibodies within her cerebrospinal fluid. A laparoscopic right partial salpingectomy and an oophorectomy were performed on day 25, after the symptoms developed further. Histopathology confirmed the presence of a mature teratoma within the right ovary. After surgery, she returned to her baseline mental status, with no further complications.

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Anti-N-methyl-D-aspartate (NMDA) encephalitis is a rare paraneoplastic syndrome that has been associated with ovarian teratomas (1). This variant of limbic encephalitis primarily affects young women and primarily manifests as psychosis, altered personality, memory deficits, encephalopathy, and/or seizures. Autonomic dysregulation and decreased consciousness, frequently requiring intubation and prolonged hospital stays, may also occur (2). In the presence of psychiatric symptoms, however, a diagnosis is often delayed (3), complicating the disease course. The management of anti-NMDA encephalitis includes supportive therapy as well as plasmapheresis, corticosteroids, rituximab, and the excision of the tumor (3). Tumor excision has been shown to improve patient outcome and greatly decrease the rate of recurrence. In this case report, we describe the case of a 22-year-old female who presented with the acute onset of psychotic symptoms and who was later diagnosed with anti-NMDA encephalitis caused by an ovarian teratoma.

Case Report

A 22-year-old female presented at our institution with altered mental status and a witnessed seizure. The patient's mother reported disorganized behavior, insomnia, inappropriate laughter, and anorexia, occurring for the past 2 weeks. At first, the patient was evaluated at a different institution and treated with antidepressants and antibiotics. Due to the development of her seizures and persistent psychiatric symptoms, she was brought to our institution for further evaluation. The patient had been afebrile during this period, as indicated by the history provided by her mother. She had a past medical history of major depressive disorder, allergic rhinitis, and recurrent urinary tract infections. Upon admission, the patient had no febrile episodes, and her vital signs were stable. In the initial evaluation by the neurology service, autoimmune encephalitis was suspected as a possible diagnosis Cerebrospinal fluid showed pleocytosis as well as increased protein concentration; for this reason, an autoimmune workup was initiated. Chest and abdominopelvic computed tomography (CT) scans showed no evidence of pathology, ruling out most common malignancies associated with autoimmune encephalitis. During her admission, the patient received high-dose steroids. However, upon initiation of an intravenous immunoglobulin (IVIG) infusion, she developed refractory status epilepticus and had to be intubated (9 days). Upon the improvement of her condition, an endovaginal ultrasound was performed to rule out ovarian teratoma, which has been related to anti-NMDA encephalitis in the literature. The endovaginal ultrasound was remarkable for a right-sided, small (1.25 cm, largest dimension) atypical intraovarian dermoid cyst/teratoma, which prompted the diagnostic consideration of limbic encephalitis caused by a paraneoplastic disorder due to the ovarian teratoma. Results showed positive anti-NMDA within the cerebrospinal fluid, specifically targeting anti-NR1. According to our literature review, the appropriate management for this case was to perform a resection of the teratoma, even though it was very small, as doing so would speed up improvement and decrease the chances

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of both a relapse and permanent brain damage (2). The patient and family were given this recommendation. A laparoscopic right partial salpingectomy and oophorectomy with the removal of the teratoma were performed. Histopathology confirmed the presence of a mature teratoma of the ovary. Posterior to her surgery, she returned to her baseline mental status and recovered well.

Discussion

Anti-NMDA receptor encephalitis is an autoimmune encephalitis that has frequently been associated with an underlying neoplasm that is usually benign in nature. In young women, this neoplasm is frequently an ovarian teratoma (2). It is important to note that this paraneoplastic syndrome presents with predominantly psychiatric symptoms, which may delay the correct diagnosis, especially in patients with a prior psychiatric diagnosis (4). Ovarian teratomas associated with anti-NMDA receptor encephalitis are usually unilateral with a mean diameter of 6 cm (5). The clinical features of anti-NMDA receptor encephalitis, which may include hallucinations, psychosis, a decreased level of consciousness, hypoventilation, and hyperthermia, are severe and may be life-threatening (1). However, nearly 80% of its patients will recover neurologically after first-line therapy: immunotherapy and tumor removal (6). Immunotherapy, such as what was employed in our patient, may include IVIG, corticosteroids, and plasma exchange (1). Tumor removal in the first 4 months after the development of symptoms has been shown to provide better outcomes as well as lower rates of relapse (3). Though these ovarian teratomas are usually unilateral, about 15% may be bilateral, with the failure of the bilateral tumor excision being a cause for a recurrence of symptoms (2). For this reason, a thorough evaluation including CT scans, magnetic resonance imaging, and endovaginal ultrasound is indicated for patients suspected to have anti-NMDA receptor encephalitis. An accurate diagnosis is important in order to start therapy as early as possible, because even in severe cases, patients usually improve after intensive care support, immunotherapy, and prolonged hospitalization (in the care of a multidisciplinary team) (7).

Resumen

Este es el caso de una mujer de 22 años que llegó a nuestra institución tras presentar insomnio refractario, comportamiento desorganizado, risa inapropiada y anorexia. Al ingreso, el examen físico reveló mutismo, irritabilidad y alucinaciones visuales. Se excluyeron causas infecciosas, metabólicas y otras causas alternativas para los síntomas que presentaba. Los resultados de la resonancia magnética cerebral y de la tomografía computarizada torácica y abdominopélvica no mostraron evidencia de patología. Debido a la alta sospecha clínica de encefalitis paraneoplásica, se inició tratamiento con dosis altas de esteroides intravenosos (IV) e inmunoglobulinas IV. Se realizó una ecografía endovaginal, que reveló un pequeño quiste dermoide intraovárico atípico. Las pruebas de laboratorio de la paciente dieron positivo para anticuerpos anti-N-metil-D-aspartato en el líquido cefalorraquídeo. Se le practicó una salpingectomía parcial derecha laparoscópica y una ooforectomía el día 25, después de que los síntomas se agravaran. La histopatología confirmó la presencia de un teratoma maduro en el ovario derecho. Tras la intervención, recuperó su estado mental basal, sin más complicaciones.

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