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Ovarian Teratomas and Anti-*N*-Methyl-D-Aspartate Receptor Encephalitis: Why Sonography First?

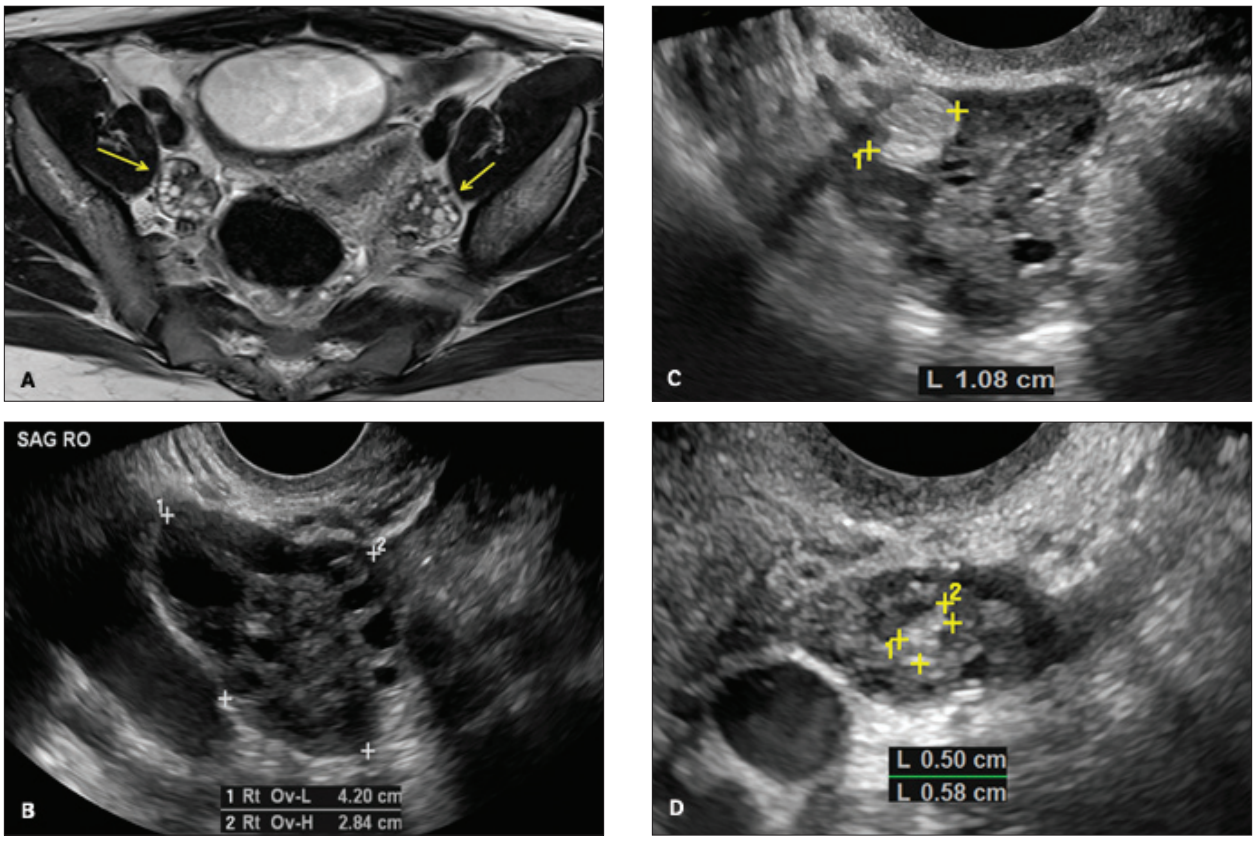
Ovarian teratomas are the most common germ cell tumors and are derived from at least 2 germ cell layers. They typically contain ectoderm (skin or brain), mesoderm (muscle or fat), and endoderm (mucinous or ciliated endothelium).[1] Anti-*N*-methyl-D-aspartate (anti-NMDA) receptor encephalitis is an autoimmune encephalitis associated with a paraneoplastic process in the setting of this type of ovarian disease. Anti-NMDA receptor encephalitis typically affects young women, with 80% of all cases being reported in women with an average age of 23 years at presentation. In a review of 100 patients with anti-NMDA receptor encephalitis, Dalmau et al[2] found that up to 54% of women had an identifiable tumor had an ovarian teratoma containing neural tissue. Patients with this condition typically present with auditory hallucinations, confusion, and neurologic deficits and can progress to autonomic instability, seizures, and unresponsiveness, leading to death. Treatment usually involves removal of the ovary with the suspected lesion, plasmapheresis, intravenous immunoglobulin, or a combination thereof. In 75% of cases, patients will have substantial improvement in their symptoms after first-line therapy; 25% of cases have persistent severe deficits or lead to death.[3–5]

At our institution, a 25-year-old woman presented with confusion and an altered mental status that progressed to agitation, delirium, and a catatonic state over the course of 1 month. She was found to have anti-NMDA receptor antibodies in her cerebrospinal fluid. During this admission, she initially underwent positron emission tomography–computed tomography and pelvic magnetic resonance imaging (MRI) with gadobutrol, in which no ovarian disease was identified (Figure 1A). Electroencephalographic findings were concerning for encephalopathy, and she received a 5-day course of intravenous solumedrol and intravenous immunoglobulins with no improvement. After receiving these medications, her condition deteriorated, and she developed malignant catatonia and dysautonomia, requiring transfer to the intensive care unit. Transvaginal pelvic sonography was subsequently performed, revealing normal-size ovaries with suspicious lesions on each. The right ovary contained a 13 × 11 × 10-mm echogenic and cystic mass. The left ovary contained a 6 × 5 × 5 mm echogenic area. The sonographic findings were consistent with bilateral ovarian teratomas (dermoids; Figure 1 B–D).

The patient ultimately underwent a laparoscopic bilateral salpingo-oophorectomy. The ovaries were normal in size on direct visualization, and no external contour abnormalities were seen. Final pathologic analysis confirmed an 8-mm mature cystic teratoma on the right ovary and a 5-mm epidermoid cyst on the left ovary. One week after surgery, the patient's clinical status showed remarkable improvement. She was alert and responsive, was able to communicate appropriately, and was ambulating. She was still confused at times and reported no memory of any events that occurred during her hospitalization. Two weeks after her surgery, she was discharged to a rehabilitation facility with short-term memory deficits but with the expectation of a full neurologic recovery.

This case highlights the heightened clinical awareness that is critical for making the diagnosis. It also emphasizes why transvaginal sonography should be the first-choice modality to evaluate the ovaries for teratomas, as they are readily identified with sonography due to their distinct imaging characteristics.[6,7] Most often, they will have an intensely echogenic component characterized by fat and sebaceous material. Strandy linear material represents hair and often originates from an echogenic area with acoustic shadowing within the cyst, known as a "Rokitansky nodule." Less commonly, teratomas may be predominantly cystic.[7] Forgoing transvaginal sonography as the initial imaging modality, on the basis of the patient's neuropsychiatric status, exposes her to unnecessary radiation at a substantial cost. Although computed tomography and MRI may be of adjunctive use, cases of "microteratomas" not previously identified by any imaging modalities or even by ovarian biopsy have been reported.[8–10] In cases in which imaging fails to identify a teratoma, some investigators advocate for bilateral oophorectomy, especially if patients do not respond

Figure 1. Ovarian teratomas and anti-NMDA receptor encephalitis in a 25-year-old woman. **A**, T2-weighted MRI showing the right and left ovaries (yellow arrows), which are normal in size and location. **B**, Coronal transvaginal sonogram of the right ovary obtained during hospitalization showing an echogenic focus characteristic of a teratoma. **C**, Sagittal view of the right ovary showing a cystic component within the echogenic mass (calipers). **D**, Sagittal view of the left ovary showing an echogenic focus measuring 5 mm, suggestive of a small teratoma and unchanged from the previous outside scan.



to immunotherapy.[9,10] Removal of both ovaries would render these young women sterile, lead them into immediate surgical menopause, and expose them to the well-known health risks of coronary artery disease and osteoporosis associated with menopause at such a young age; however, this approach may be their best opportunity for survival.[10] Hence, pelvic sonography should be the first-line imaging modality to assess for teratomas in a female patient with a diagnosis of anti-NMDA receptor encephalitis.

Roberto J. Vargas, MD, Huma Farid, MD, Robin P. Goldenson, MD, MPH, Alexandra H. Fairchild, MD, Benjamin J. Dorton, MD, Bryann S. Bromley, MD

Department of Obstetrics and Gynecology
Brigham and Women's Hospital and
Massachusetts General Hospital
Boston, Massachusetts USA (R.J.V., H.F., B.J.D., B.S.B.)
Department of Radiology
Brigham and Women's Hospital
Boston, Massachusetts USA (R.P.G., A.H.F., B.S.B.)
Harvard Medical School
Boston, Massachusetts USA (R.J.V., H.F., R.P.G., A.H.F., B.J.D., B.S.B.)

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