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Delayed Opsoclonus–Myoclonus Syndrome After Ovarian Teratoma Resection

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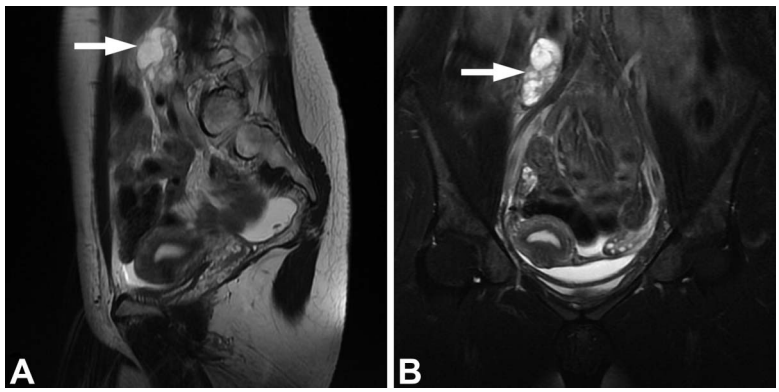

FIG. 1. Pelvic MRI. T2 sagittal (**A**) and coronal (**B**) MRI of the pelvis demonstrate a large, complex, cystic adnexal mass with septations (*arrow*) in the right paravertebral lower abdomen, which was found to be mature teratoma on pathologic examination.

Abstract: Opsoclonus–myoclonus syndrome (OMS) is a rare syndrome characterized by opsoclonus, which is irregular, spontaneous, multivectorial saccadic eye movements, along with diffuse or focal myoclonus and sometimes ataxia. OMS is associated with paraneoplastic etiologies in 20%–40% of cases, with small-cell lung and breast cancers the most common associated primary neoplasms in adults, whereas neuroblastoma is more common in children and ovarian teratoma may occur in women younger than 30 years. Onconeural antibodies are often not identified. In existing literature, paraneoplastic OMS precedes identification of the neoplasm, and neurological recovery depends on treatment of the underlying cancer. We describe a 27-year-old woman with the delayed onset of OMS one month after resection of ovarian teratoma, likely due to immune trigger from antigen exposure at the time of resection. She was treated with intravenous methylprednisolone, immunoglobulins, and eventually rituximab with resolution of her symptoms. Identification of OMS after tumor resection and prompt immunotherapy are critical for neurologic recovery. At 30-month follow-up, this patient had not experienced recurrence of OMS.

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A 27-year-old woman presented to hospital with several months of early satiety and abdominal pain. Ultrasound and MRI demonstrated a right adnexal mass (Fig. 1) which was resected successfully, revealing mature teratoma. Medical, family, and medication histories were otherwise noncontributory. One month later, she returned to the emergency department with acutely worsening nausea, dizziness, and unsteady gait. Examination revealed a visual acuity of 20/20 in each eye with equal and reactive pupils and was most notable for opsoclonus and saccadic pursuits (see **Supplemental Digital Content**, Video E1, http://links.lww.com/WNO/A504), postural tremor, action myoclonus, and wide-based gait. MRI of the brain and infectious and autoimmune serology results were unremarkable. Cerebrospinal fluid (CSF) analysis showed normal glucose, protein, and no cells or oligoclonal bands. Onconeural antibodies were negative, including against N-methyl-D-aspartate receptors. Repeat pelvic imaging

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The authors report no conflicts of interest.

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and positron emission tomography demonstrated no residual or additional tumor. Given recent ovarian teratoma, paraneoplastic Opsoclonus–myoclonus syndrome (OMS) was diagnosed, and her symptoms rapidly resolved after 3 days of intravenous methylprednisolone 1 gm daily and intravenous immunoglobulin (IVIg) 2 gm per kilogram. Her symptoms relapsed after one month, and once again, imaging and CSF remained unremarkable. She was treated with a second course of intravenous methylprednisolone and IVIg as well as induction rituximab 2 doses of 1 gm each with subsequent doses after 6 and 12 months, with no further symptoms. At 30-month follow-up, she had not experienced recurrence of opsoclonus or myoclonus and has not required any further immunotherapy.

OMS—also known as "dancing eyes" syndrome—is a rare syndrome characterized by opsoclonus, which is irregular, spontaneous, multivectorial saccadic eye movements (see **Supplemental Digital Content**, Video E1, http://links.lww.com/WNO/A504), along with diffuse or focal myoclonus and sometimes ataxia (1). The exact pathophysiology remains unknown; however, it has been postulated to involve loss of inhibitory signals either from Purkinje cells in the cerebellar vermis (2) or from omnipause cells which normally inhibit burst neurons responsible for generating saccades (3). OMS etiology may be idiopathic, autoimmune, parainfectious, or paraneoplastic, with both humoral and cell-mediated immune-mediated mechanisms suspected (4–6). A wide variety of viral or bacterial pathogens have been implicated, including the severe acute respiratory syndrome coronavirus 2 (7). Paraneoplastic etiology has been identified in 20%–40% of patients (5,6). Small-cell lung and breast cancers are the most commonly associated primary neoplasms in adults, whereas neuroblastoma is more common in children and ovarian teratoma may occur in women younger than 30 years. (4–6). Onconeural antibodies are often not identified (4–6). Management relies on early detection and treatment of the underlying neoplasm if present, and further clinical response with intravenous corticosteroids, IVIg, cyclophosphamide, or rituximab (5,6). Symptomatic improvement is reported with clonazepam (8) and topiramate (9).

In existing literature, paraneoplastic OMS precedes identification of the neoplasm, and neurological recovery depends on treatment of the underlying cancer (4–6). However, this case uniquely illustrates a delayed onset of OMS one month after resection of ovarian teratoma, likely due to immune trigger from antigen exposure at the time of resection. Identification of OMS after tumor resection and prompt immunotherapy are critical for neurologic recovery.

STATEMENT OF AUTHORSHIP
Category 1: a. Conception and design: T. Chen; b. Acquisition of data: T. Chen; c. Analysis and interpretation of data: T. Chen. Category 2: a. Drafting the manuscript: T. Chen and A. A. Jones; b. Revising it for intellectual content: T. Chen and A. A. Jones. Category 3: a. Final approval of the completed manuscript: T. Chen and A. A. Jones.

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