

The most remarkable finding in our case is that the neurological symptoms were a paraneoplastic manifestation of an occult cancer. Although we strongly suspected that the patient had a testicular tumor, we were unable to demonstrate the tumor before the orchiectomy. The reasons for orchiectomy were relentless progression of neurological symptoms, young age of the patient, detection of anti-Ma2 antibodies, and detection of microcalcifications in the follow-up ultrasound. Testicular microcalcifications have been reported with a variable incidence (from 0.6% to 18.1%),<sup>9,10</sup> among individuals undergoing ultrasound and associates with development of germ-cell tumors in 40% to 45% of patients.<sup>9</sup>

IGCN is considered a precursor of most testicular germ-cell tumors. It consists of enlarged cells with clear cytoplasm that are aligned along the basal portion of the seminiferous tubules.<sup>11</sup> Immunostaining discloses PLAP-positive cells in most of the patients with IGCN. It should be noted that nonneoplastic spermatogenic cells are almost always PLAP-negative. Therefore, the identification of dysplastic PLAP-positive cells in seminiferous tubules, as in our patient, is confirmatory to the diagnosis of IGCN.

The prognosis of anti-Ma2-associated encephalitis depends on prompt treatment of the tumor, and some patients appear to respond to immunosuppressants (corticosteroids or intravenous immunoglobulin).<sup>1</sup> Factors associated with improvement or stabilization include male gender, young age, testicular tumor with complete response to treatment, absence of anti-Ma1 antibodies, and limited central nervous system involvement.<sup>1</sup> In our case, the neurological symptoms showed a rapidly progressive course, and the diagnosis was established several months after the symptom presentation. Because there was severe involvement of the CNS and the affected brain regions were already atrophic at the time of orchiectomy, it resulted in only stabilization of symptoms, but not improvement. We conclude that anti-Ma2 encephalitis should be considered in patients with subacutely progressive severe hypokinesia. If the patient is a young man without evidence of a tumor, a microscopic IGCN should be strongly considered. In these patients, orchiectomy can be the only way to reveal the tumor at very early stages. Early diagnosis and treatment should be encouraged because of its beneficial effects when compared with other paraneoplastic neurological syndromes.<sup>1,3</sup>

#### LEGENDS TO THE VIDEO

**Segment 1.** The muscle tone of the neck and limbs was increased and associated with severe rigidity and spasticity. Intermittent dystonic postures were also observed.

**Segment 2.** The patient was able to answer questions correctly by protruding his tongue. Note that the tongue movements are extremely slow.

**Segment 3.** The patient was asked to raise his left arm; note that the movement is very slow. It took a few minutes to complete this movement.

**Segment 4.** This segment shows an episode of diaphoresis with tachycardia and tachypnea.

#### REFERENCES

1. Dalmau J, Graus F, Villarejo A, et al. Clinical analysis of anti-Ma2-associated encephalitis. *Brain* 2004;127:1831–1844.
2. Rosenfeld MR, Eichen JG, Wade DF, Posner JB, Dalmau J. Molecular and clinical diversity in paraneoplastic immunity to Ma proteins. *Ann Neurol* 2001;50:339–348.
3. Voltz R, Gultekin SH, Rosenfeld MR, et al. A serologic marker of paraneoplastic limbic and brain-stem encephalitis in patients with testicular cancer. *N Engl J Med* 1999;340:1788–1795.
4. Mathew RM, Yamamoto T, Nakamura K, Dropcho E, Tsuji S, Dalmau J. Orchiectomy for suspected microscopic tumor in patients with paraneoplastic anti-Ma2 encephalitis. In: *Contemporary Clinical Issues and Case Studies, Plenary Session: 58th Annual Meeting AAN, San Diego, CA, April 5, 2006*. Available at [http://www.abstracts2view.com/aan/view.php?nu=AAN06L\\_PL01.001](http://www.abstracts2view.com/aan/view.php?nu=AAN06L_PL01.001)
5. Castle J, Sakonju A, Dalmau J, Newman-Toker DE. Anti-Ma2-associated encephalitis with normal FDG-PET: a case of pseudo-Whipple's disease. *Nat Clin Pract Neurol* 2006;2:566–572.
6. Feve AP, Fenelon G, Wallays C, Remy P, Guillard A. Axial motor disturbances after hypoxic lesions of the globus pallidus. *Mov Disord* 1993;8:321–326.
7. Bhatia KP, Marsden CD. The behavioral and motor consequences of focal lesions of the basal ganglia in man. *Brain* 1994;117:859–876.
8. Aizawa H, Kwak S, Shimizu T, et al. A case of adult onset pure pallidal degeneration. II. Clinical manifestations and neuropathological observations. *J Neurol Sci* 1991;102:76–82.
9. Hobarth K, Susani M, Szabo N, Kratzik C. Incidence of testicular microlithiasis. *Urology* 1992;40:464–467.
10. Middleton WD, Teefey SA, Santillan CS. Testicular microlithiasis: prospective analysis of prevalence and associated tumor. *Radiology* 2002;224:425–428.
11. Montironi R. Intratubular germ cell neoplasia of the testis: testicular intraepithelial neoplasia. *Eur Urol* 2002;41:651–654.