

Appendix e-1

Information on the 4 patients who did not respond to treatment and had a tumor and/or additional onconeuroal antibodies

The patient with SOX1 antibodies had SCLC and presented with LE. His symptoms stabilized with steroids and chemotherapy but he died 2 years later due to tumor progression. The patient with amphiphysin antibodies presented with diffuse encephalitis with limbic dysfunction, insomnia, and ataxia. He did not respond to steroids; several months later a mass was found radiologically in the lung and he died due to pulmonary embolism before tumor diagnosis was histologically confirmed; no autopsy was performed. One patient with CRMP5 antibodies had a malignant thymoma that was treated with chemotherapy and radiation. Few months later he presented with LE and status epilepticus but died shortly after the first cycle of IVIg and steroids due to pulmonary embolism, therefore, the effects of immunotherapy were not assessable. The other patient with CRMP5 antibodies presented with memory deficits, psychiatric symptoms, and optic neuropathy. He received steroids and rituximab; the psychiatric symptoms improved, but he developed ataxia and cachexia; no tumor has been identified to date.