

Approximately 25% of patients with herpes simplex encephalitis develop a form of autoimmune encephalitis that usually is associated with abnormal movements (choreoathetosis after herpes simplex encephalitis) in children and with cognitive and psychiatric symptoms in adults. This disorder develops a few weeks after the viral infection has resolved, is associated with new synthesis of antibodies against the NMDA receptor and other neuronal cell surface proteins, and is usually less responsive to immunotherapy than anti-NMDA receptor encephalitis (idiopathic or teratoma-associated).

Encephalitis with α -amino-3-hydroxy-5-methylisoxazole-4-propionic acid (AMPA) receptor antibodies affects middle-aged women, who develop acute limbic dysfunction or, less frequently, prominent psychiatric symptoms; 70% of patients have an underlying tumor in the lung, breast, or thymus (Fig. 94-4B). In about 50% of cases, the brain MRI shows typical features of limbic encephalitis (similar to Fig. 94-2A). Neurologic relapses may occur; these also respond to immunotherapy and are not necessarily associated with tumor recurrence.

Encephalitis with GluK2 antibodies can affect children and adults and is associated with rapidly progressive encephalopathy with cerebellar ataxia or cerebellitis. Symptoms of encephalopathy may include impairment of memory and level of consciousness and motor alterations such as dyskinesias, choreoathetosis, bradykinesia, and spastic paraparesis. Some patients develop intracranial hypertension. In one patient, the symptoms were associated with teratoma.

Encephalitis with γ -aminobutyric acid type A (GABA_A) receptor antibodies may affect children and adults and is associated with prominent seizures and status epilepticus often requiring a pharmacologically induced coma. In approximately 80% of patients, the brain MRI shows multifocal, asynchronous, cortical-subcortical T2/FLAIR abnormalities predominantly involving temporal and frontal lobes, but also basal ganglia and other regions (Fig. 94-2C). Most patients do not have an underlying tumor, but some, usually of Japanese ethnicity, may have thymoma.

Encephalitis with GABA_B receptor antibodies is usually associated with limbic encephalitis and seizures. In >50% of cases,