On-Line Table: Differential diagnosis of rapidly progressive dementias

Rapidly Progressive Dementias	MR Imaging Characteristics
Neurodegenerative conditions	
Variants of typically slowly progressive dementia	
Alzheimer disease	Diffuse atrophy, particularly within hippocampus, medial temporal lobes (with enlargement of temporal horns); MRS shows decreased NAA, increased myo-inositol
Frontotemporal dementia	Generalized atrophy of frontal and temporal lobes may be detected with MRI; "knife edge" sign with focal atrophy of temporal lobe
Progressive neurodegenerative dementias	
Dementia with Lewy bodies	Nonspecific global and subcortical atrophy, relative preservation of medial temporal lobe
Corticobasal degeneration	Atrophy within caudate nucleus, putamen, frontal and superior parietal cortices; T2-weighted signal increase in subcortical WM
Progressive supranuclear palsy	Severe atrophy of midbrain, pons with "hummingbird" or "penguin" sign on sagittal images; Low T2 signal within putamen with increased ADC; T2 hyperintensity of tegmentum, loss of sagittal convexity of midbrain profile
Parkinson disease with dementia	Thinning of pars compacta occasionally (T2 hyperintense band of substantia nigra); decreased FA and magnetization transfer ratio in substantia nigra
Huntington disease	Atrophy of caudate nucleus (with enlargement of frontal horns), sometimes T2 hyperintense; T2/FLAIR hypointensity within basal ganglia with iron deposition
Multiple-system atrophy	Severe atrophy within brain stem, cerebellum, putamen with putaminal hypointensity on T2 with hyperintense rim and increased ADC; pons T2/FLAIR hyperintensity, "hot cross bun" sign with decreased FA within pons and middle cerebellar peduncle
Prion-related conditions	
Creutzfeldt-Jakob disease	DWI hyperintensity within basal ganglia, thalamus, and cortex with "cortical ribboning" (usually occipital); "hockey stick" sign; DWI hyperintensity within dorsomedial thalamus and pulvinar
Infectious-disease related conditions	
Viral encephalitis	FLAIR and T2 hyperintensity within temporal (particularly, medial), insular, and inferior frontal cortices in herpes; thalami and subcortical GM in Japanese encephalitis; DWI hyperintensity/ADC reduction in same distribution acutely; restricted diffusion in insular cortex bilaterally is pathognomonic
HIV-related dementia	Periventricular T2/FLAIR hyperintensity without contrast enhancement; diffuse atrophy; MRS reductions in glutamate/glutamine and NAA
Tertiary syphilis	Meningovascular contrast-enhancement; T2 and FLAIR hyperintensities in arterial distribution
Neuroborreliosis (Lyme disease)	Nonspecific T2 hyperintense foci, cranial nerve enhancement
Subacute sclerosing panencephalitis (pediatric age group)	Periventricular T2 hyperintensities; hyperintense lesions in parietal/temporal lobes; decreased FA within white matter on DTI; MRS shows decreased NAA, increased choline, lipid, and lactate
Variant Creutzfeldt-Jakob disease	As described under prion-related conditions
Inflammatory/autoimmune conditions	
Vasculitis	Nonspecific T2-weighted and FLAIR hyperintensities; vessel wall thickening seen on MRI and MRA
Hashimoto encephalopathy/steroid-responsive encephalopathy with autoimmune thyroiditis	Nonspecific subcortical white matter abnormalities on FLAIR; contrast-enhancement of meninges
Multiple sclerosis	Diffuse atrophy; reduction in NAA in normal-appearing gray and white matter on MRS
Lupus cerebritis	Deep white matter loss, scattered white matter lesions
Neurosarcoidosis	Contrast-enhancing dural plaques; periventricular T2 hyperintensities; multiple brain lesions; leptomeningeal enhancement
Celiac-associated encephalopathy	Hyperintense T2 foci in posterior white matter, nonspecific
Neoplasm-related conditions	
Paraneoplastic autoimmune encephalitis	Mesial temporal lobe hyperintensity on FLAIR
Lymphoma, primary CNS	T1 isointense to slightly hyperintense, T2 slightly hypointense mass; homogeneous enhancement; classic "butterfly" lesion crossing corpus callosum; MRS shows increased choline, lipid, and lactate; MR perfusion shows rCBV lower than that in GBM or metastases; multiple lesions more common in AIDS, solitary more common in the immunocompetent

On-Line Table: (Continued)

Rapidly Progressive Dementias	MR Imaging Characteristics
Metastatic malignancy	Multiple small lesions at GM-WM junction, frequently edematous with T2 hyperintensity; T1 hyperintensity if hemorrhage; peripheral contrast enhancement DTI demonstrates displacement, not invasion, of white matter tracts; increased rCBV and rCBF
Lymphomatoid granulomatosis	Multifocal punctate and linear enhancing lesions; temporal lobe atrophy in some cases
Toxic/metabolic/endocrine conditions	
Metals (eg, manganese)	T1 hyperintensity within basal ganglia, particularly globus pallidus
Vitamin B_1 (thiamine) deficiency (Wernicke encephalopathy)	T2-weighted and FLAIR hyperintensities within periaqueductal regions, around third ventricle and bilateral thalami; contrast enhancement of mammillary bodies
Mitochondriopathy, MELAS	Cortical T2-based hyperintensities, corresponding DWI hyperintensity in gyriform pattern
Wilson disease	Diffuse atrophy with signal irregularities in deep gray and white matter, "panda sign"
Osmotic myelinolysis	Diffuse hyperintensity within basis pontis on T2/FLAIR, with restricted diffusion, with "trident" sign

Note:—MELAS indicates mitochondrial myopathy; FA, fractional anisotropy; GBM, glioblastoma multiforme; GM, gray matter; rCBF, relative cerebral blood flow; rCBV, relative cerebral blood volume.