

Index

abscess, brain, 186, 542, 554 aceruloplasminemia (ACP), 15–16 neuroferritinopathy vs., 8, 16 acromesomelic upper limb shortening, 569 acute cerebellitis, 153–6 acute confusional state, 116 acute disseminated encephalomyelitis (ADEM) acute cerebellitis vs., 156 acute necrotizing encephalopathy vs., 252 hemorrhagic, vs. malaria, 208 Susac syndrome vs., 104 Wernicke encephalopathy vs., 360 acute necrotizing encephalopathy, 249–53 acute hyperammonemic encephalopathy, 301–5 adrenoleukodystrophy, 40 X-linked (XLA), 264 AIDS. <i>See</i> HIV infection/AIDS alcohol abuse Marchiafava-Bignami disease, 308 Wernicke encephalopathy, 357, 361 Alexander disease, adult-onset, 46, 373–6 Alzheimer disease (AD) corticobasal syndrome, 26 hippocampal sclerosis dementia vs., 36 mammillothalamic tract degeneration vs., 588 ammonia toxicity, 304 amnesia acute-onset, after thalamic infarct, 587–8 psychogenic, 116 transient global (TGA), 113–16 amyloid β -related angiitis (ABRA), 94–5 amyloidoma, cerebral (CA), 87–91 amyloidosis, 90–1 amyotrophic lateral sclerosis (ALS), 40–1, 212 anterior cerebral artery infarction, 308 anterior inferior cerebellar artery (AICA), 656 antiangiogenic therapy. <i>See</i> bevacizumab antiepileptic drug withdrawal, 327–30 anti-Ma2-associated paraneoplastic encephalitis (AMAPE), 445–7 anti-NMDA receptor encephalitis, 223–6 aquaporin-4 (AQP4) autoantibodies, 258 arachnoid cysts, 476 arachnoid granulations, giant, 607–11 arborized pattern, schistosomiasis, 192 arteriovenous malformations (AVMs) differential diagnosis, 78, 386, 504 radiation-induced tumefactive cyst, 131–4 aspergillosis, 183–8 astroblastoma, 389–93 astrocytoma fibrillary, 398 infiltrating low-grade (ILGA), 488 low-grade, 524 pilocytic (PA), 548 pilomyxoid (PMA), 18, 548–9 pineal region, 498 protoplasmic, 395–9 <i>ATP7A</i> gene, 298 <i>ATP7B</i> gene, 342 <i>ATXN3</i> gene, 72 atypical teratoid/rhabdoid tumor, 404, 410 autonomic failure, multiple system atrophy, 21–2 autosomal dominant leukodystrophy (ADLD), adult-onset, 66 Avastin. <i>See</i> bevacizumab bag of worms appearance, 504 Balo concentric sclerosis (BCS), 245–7 basal ganglia aspergillosis, 184, 186 chronic hepatic encephalopathy, 291 cystic degeneration, 7–8 hemolytic uremic syndrome, 141–2, 144 Huntington disease, 55–7 hybrid phakomatosis, 577, 580 hyperglycemic hemiballismus and hemichorea, 281–2 multiple system atrophy, 21–2 osmotic demyelination syndrome, 371–2 phospholipase-associated neurodegeneration, 13–14 Whipple disease, 18 Wilson disease, 340, 342–3 basal ganglia calcification aceruloplasminemia vs., 16 chronic hepatic encephalopathy, 292 familial idiopathic (FIBGC), 16, 614 mineralizing microangiopathy, 613–14 bat-wing pattern, osmotic demyelination syndrome, 372 Behçet disease, 228 neurologic (NBD), 227–9, 242 rhombencephalitis, 160 Bell palsy, 152 beta-propeller protein-associated neurodegeneration (BPAN), 2, 4 bevacizumab distal recurrence of glioblastoma after, 443–4 for hippocampal radiation necrosis, 430 persistent diffusion restriction after, 494 pseudoresponse of glioblastoma to, 439–42 Bickerstaff brainstem encephalitis (BBE), 237–40, 242 biphasic myelinopathy of Grinker, 288 blastomycosis, 181–2 blepharitis, lipid proteinosis, 268 Bloch-Siemens syndrome. <i>See</i> incontinentia pigmenti BPAN. <i>See</i> beta-propeller protein-associated neurodegeneration brain atrophy corticobasal syndrome, 26 frontotemporal dementia with <i>FUS</i> mutation, 59 Machado-Joseph disease, 71–2 Menkes disease, 298 MPAN, 3–4 neuroferritinopathy, 8 phospholipase-associated neurodegeneration, 14 progressive supranuclear palsy, 27–8 brainstem. <i>See also</i> medulla oblongata; midbrain; pons adult-onset Alexander disease, 373, 376 blastomycosis, 181–2 encephalitis. <i>See</i> rhombencephalitis glioma, 182, 272 Machado-Joseph disease, 69, 72 metronidazole-induced encephalopathy, 322–4 neuromyelitis optica spectrum disorder, 258 progressive ataxia and palatal tremor, 43, 46 branch retinal artery occlusion (BRAO), 105 bright claustral sign, Wilson disease, 343 <i>C19orf12</i> gene, 4 CADASIL, 107–11

Index

- calcification
 astroblastoma, 389, 392
 basal ganglia. *See* basal ganglia
 calcification
 brain tumors with, 90, 268
 cerebral amyloidoma, 87, 90
 deep gray nuclei, 16
 dermoid cysts, 484
 intracranial chondroma, 458
 lipid proteinosis, 267–8
 meningioangiomas, 383, 386
 mineralizing microangiopathy, 613–14
 rosette-forming glioneuronal tumor, 382
 T1-weighted hyperintensity, 84
 third ventricle
 craniopharyngioma, 461, 464
 calcineurin inhibitor-mediated
 bilateral limbic injury, 599–602
 capillary telangiectasias (CT), 128
 carbon monoxide poisoning, 10, 342
 delayed encephalopathy, 285–8
 cardiac arrest, with global hypoxia, 84
 caudate nuclei
 chronic infarcts, 56
 frontotemporal dementia with *FUS* mutation, 59
 Huntington disease, 55–7
 Wilson disease, 342
 cavernous malformations
 cavernous sinus, 530
 developmental venous anomaly and, 128
 meningioangiomas vs., 386
 cavernous sinus hemangioma, 529–30
 cephalocele, 148
 cerebellar ataxia
 Erdheim-Chester disease, 623
 idiopathic late-onset (ILOCA), 20
 cerebellar atrophy, 14
 cerebrotendinous xanthomatosis, 49
 MPAN, 3–4
 multiple system atrophy, 19–20
 phospholipase-associated neurodegeneration, 13–14
 progressive supranuclear palsy, 27–8
 cerebellar liponeurocytoma, 421–4
 cerebellar peduncles. *See also* middle cerebellar peduncles
 LBSL, 64, 66
 Machado-Joseph disease, 69, 72
 primary lateral sclerosis, 38, 40
 cerebellitis, acute, 153–6
 cerebellum
 cerebrotendinous xanthomatosis, 49, 52
 rosette-forming glioneuronal tumor, 381–2
 cerebral amyloid angiopathy (CAA)
 related inflammation (CAA-RI), 94–5
 without inflammation, 94
 cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL), 107–11
 cerebral proliferative angiopathy (CPA), 78–9
 cerebral venous thrombosis
 bilateral thrombosis of internal cerebral vein, 135–9
 differential diagnosis, 128, 252, 278, 610
 cerebrotendinous xanthomatosis (CTX), 49–52, 66, 72
 ceruloplasmin
 gene mutations, 16
 low serum, 16, 339, 342
 cervical compressive myelopathy, 40
 cervical spinal cord
 adult-onset Alexander disease, 376
 cerebrotendinous xanthomatosis, 49, 52
 Chagas disease, 199–203
 child abuse, 298
 childhood ataxia with central nervous system hypomyelination. *See* vanishing white matter (VWM) disease
 chondroma
 dural convexity, 455–8
 meningioma vs., 452, 458
 chondrosarcoma, 458, 530
 chordoid glioma, 464, 548
 chordoma, 530
 choroid plexus carcinoma, 560
 choroid plexus papilloma, temporal horn, 560
 CLIPPERS (chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids), 241–3
 colloid cysts, 464
 colonic adenocarcinoma, mucinous, 480
 congenital diseases manifesting in adults, 566
 copper
 deficiency, 366
 deposition in brain, 342
 metabolic/transport defects, 298, 342
 corpus callosum (CC)
 hereditary spastic paraplegia with hypoplastic, 566
 isolated infarction, 308
 Marchiafava-Bignami disease, 307–9
 neuromyelitis optica spectrum disorder, 258–9
 partial agenesis, 308
 phospholipase-associated neurodegeneration, 14
 splenium. *See* splenium of corpus callosum
 Susac syndrome, 103–5
 cortical dysplasia, pericallosal and left frontal surface lipoma with, 512–15
 cortical laminar necrosis, 304
 corticobasal degeneration (CBD), 26, 60
 corticobasal syndrome (CBS), 25–6, 28, 60
 corticospinal tract
 HTLV-1-related tropical spastic paraparesis, 212
 multiple system atrophy, 21
 primary lateral sclerosis, 38, 40
 craniectomy, sinking skin flap syndrome after, 603–6
 craniopharyngioma, third ventricle, 461–5
 Creutzfeldt-Jakob disease (CJD), 26, 304
 variant, 360
 cutis verticis gyrata, 643–6
 cyclosporine A, 602
 cysticercosis. *See* neurocysticercosis
 cysts/cystic lesions
 astroblastoma, 389, 392
 brain tumors, 392
 dermoid cysts. *See* dermoid cysts
 epidermoid cysts. *See* epidermoid cysts
 extraventricular neurocytoma, 415, 418
 glioblastoma with, 542
 microcystic meningioma, 449, 452–3
 neurocysticercosis, 213–14
 neuroferritinopathy, 7–8
 pilomyxoid astrocytoma, 545, 548
 protoplasmic astrocytoma, 398
 radiation-induced tumefactive cyst, 131–4
 supratentorial ependymoma, 517, 520
 third ventricle
 craniopharyngioma, 461, 464
 vanishing white matter disease, 263–4
 cytomegalovirus encephalitis, 360
 dark dentate disease, 46
DARS2 gene, 66
 delayed ischemic hyperintensity (DIH), 84–5
 demyelination
 Balo concentric sclerosis, 246
 delayed carbon monoxide poisoning, 288
 Marchiafava-Bignami disease, 309
 osmotic demyelination syndrome (ODS), 272, 369–72
 progressive multifocal leukoencephalopathy, 166
 subacute combined degeneration, 366
 tumefactive, 272
 dentate nuclei
 cerebrotendinous xanthomatosis, 49, 52
 Erdheim-Chester disease, 620, 622
 Machado-Joseph disease, 69, 72
 dentatorubral-pallidoluysian atrophy (DRPLA), 72, 584
 dentato-rubro-olivary pathway. *See* Guillain-Mollaret triangle
 dermoid cysts, 414, 484
 intra-axial, 481–4
 lipoma vs., 514
 malignant transformation, 411–14
 ruptured, 484, 536
 white epidermoid cysts vs., 476
 desmoplastic medulloblastoma, multifocal, 407–10
 developmental venous anomalies (DVA), complications, 125–9
 diabetes mellitus
 aceruloplasminemia, 15–16
 hyperglycemic hemiballismus and hemichorea, 84, 281–3
 DIDMOAD (diabetes insipidus, diabetes mellitus, optic atrophy, and deafness), 351–4
 diffuse axonal injury (DAI), 122
 drop metastases
 pinealoblastoma with, 498–9
 renal cell carcinoma with, 503–4
 dural convexity chondroma, 455–8
 dural tail sign
 hemangiopericytoma, 510
 meningioma, 452

Index

- dural venous sinuses
 arachnoid granulations, 610
 idiopathic intracranial hypertension, 595–6
 dysembryoplastic neuroepithelial tumor (DNET), 524
 differential diagnosis, 268, 398
 multifocal, 524
- ear of the lynx sign, 564, 566
- edema, cerebral. *See also* vasogenic edema
 hemolytic uremic syndrome, 144
- internal cerebral vein
 thrombosis, 136, 138
- meningoangiomas, 383, 386
- metronidazole-induced encephalopathy, 324
- microcystic meningioma, 453
- empty sella, 594, 596
- en coup de sabre appearance, 639–40
- encephalitis
 brainstem. *See* rhombencephalitis
 paraneoplastic. *See* paraneoplastic encephalitis syndromes
 Susac syndrome vs., 104
 viral. *See* viral encephalitis
- encephalocraniocutaneous lipomatosis (ECCL), 514
- eosinophilic granuloma, calvarial, 148
- ependymomas, 520
 supratentorial, 392, 520
- epidermoid cysts
 giant arachnoid granulation vs., 610
 intraparenchymal, 467–70
 white, 473–6
- Epstein-Barr virus (EBV)
 encephalitis, 160
- Erdheim-Chester disease, 619–23
- extracellular matrix protein 1 (ECM1) gene, 268
- eye-of-the-tiger sign
 absence, 4
 differential diagnosis, 10
 neuroferritinopathy, 8
 PKAN, 10
 Wilson disease, 342
- face of the giant panda and her cub sign, 343
- facial appearance
 lipid proteinosis, 267–8
 Parry-Romberg syndrome, 637, 640
 trigeminal trophic syndrome, 647, 649–50
- familial idiopathic basal ganglia calcification (FIBGC), 16, 614
- fat deposits
 cerebellar liponeurocytoma, 421, 424
 dermoid cyst, 412, 414, 484
 intratumoral, differentiation, 414, 424
 lipoma, 512, 514
 ruptured dermoid cyst, 484, 533, 536
- fat embolism, cerebral, 119–23
- ferritin
 light chain gene mutations, 8
 serum, 7, 16, 345, 348
- fibrillary astrocytoma, 398
- fibrinoid leukodystrophy, 376
- filling-in enhancement, cavernous sinus
 hemangioma, 528, 530
- flame appearance. *See* ear of the lynx sign
- FMRI* gene, 584
- fourth ventricle, rosette-forming glioneuronal tumor, 381–2
- fragile X-associated tremor/ataxia syndrome (FXTAS), 72, 581–5
- Friedrich ataxia, 354
- frontal lobe
 corticobasal syndrome, 26
 frontotemporal dementia with *FUS* mutation, 59
 Whipple disease, 18
- frontotemporal dementia (FTD), 28, 60
 behavioral variant (bv-FTD), 60
- hippocampal sclerosis
 dementia vs., 36
 with *FUS* gene mutation (FTD-FUS), 59–61
 with motor neuron disease (FTD-MND), 60
- frontotemporal lobar degeneration (FTLD), 60
 corticobasal syndrome, 26
 right temporal variant, 32
- frontotemporal lobar degeneration (FTLD)-FUS, 59–61
- frontotemporal lobar degeneration (FTLD)-tau, 60
- frontotemporal lobar degeneration (FTLD)-TDP-43, 60
- frontotemporal lobar degeneration (FTLD)-U, 60
- fungal abscess, 542, 554
- FUS* gene mutation, frontotemporal dementia associated with (FTD-FUS), 59–61
- fused in sarcoma (FUS) protein, 60
- ganglioglioma, 268, 418, 524
- germ cell tumors (GCT)
 hypothalamic, 464
 pineal region, 498
 testicular, paraneoplastic encephalitis, 445–7
- glial fibrillary acidic protein (GFAP) gene, 376
- glioblastoma (GBM), 542
 cystic, 542
 differential diagnosis, 134, 398, 554
 distal recurrence after antiangiogenic therapy, 443–4
 persistent diffusion restriction after bevacizumab, 494
 pseudoprogression after chemoradiation, 433–7
 pseudoresponse after antiangiogenic therapy, 439–42
- glioma
 angiocentric, 524
 brainstem, 182, 272
 chordoid, 464, 548
 gliomatosis cerebri, 488
- globus pallidus
 BPAN, 2
 MPAN, 4
 neuroferritinopathy, 7–8
 PKAN, 9–10
- glutaric aciduria type 1, 298
- granulomas
Aspergillus, 187
 neurosarcoidosis, 234
 tuberculosis, 193, 196
- Guillain-Mollaret triangle, 627–8
- hypertrophic olivary degeneration, 628
- progressive ataxia and palatal tremor, 46
- gumma, syphilitic cerebral, 175–9
- hair-curled pattern, epidermoid cyst, 470
- headache
 idiopathic intracranial hypertension, 593, 596
 postural, 589–90
 recurrent acute thunderclap, 97, 100
- hearing impairment, Susac syndrome, 105
- hemangioblastoma (HGBL), 504
- hemangiomas. *See also* venous malformations
 cavernous. *See* cavernous malformations
 cranial osseous, 148
- hemangiopericytoma, intraventricular, 510
- hemiballismus and hemichorea, 282
- hyperglycemic (HHH), 84, 281–3
- hemifacial atrophy, Parry-Romberg syndrome, 637, 640
- hemifacial microsomia, congenital, 640
- hemochromatosis, hereditary, 345–8
- hemolytic uremic syndrome, 141–4
- hemorrhage, intracranial. *See also* microhemorrhages; subarachnoid hemorrhage
 cerebral malaria, 208
 chronic hepatic leukoencephalopathy, 288
 developmental venous anomalies, 128
 hypertensive, 278
 posterior reversible encephalopathy syndrome, 275–9
- hemorrhagic infarction
 internal cerebral vein thrombosis, 138
 T1-weighted hyperintensity, 84
- hepatic encephalopathy, 292
 chronic, 282, 292, 342
- hepatic leukoencephalopathy, chronic, 288
- hepatolenticular degeneration. *See* Wilson disease
- hereditary spastic paraplegia (HSP), 566
 differential diagnosis, 40, 566
 with hypoplastic corpus callosum (HSP-HCC), 566
- herpes simplex virus (HSV)
 acute cerebellitis, 153–6
 encephalitis, 104, 160
- herpes virus type 6 (HHV-6)
 encephalopathy, 602
- herpes zoster oticus. *See* Ramsay Hunt syndrome
- Herpesviridae, 156
- hippocampal sclerosis
 other associated neuropathologies, 36
 pure, with dementia, 33–6
- hippocampus
 anti-NMDA receptor encephalitis, 223, 225
 calcineurin inhibitor-mediated injury, 599–602
 radiation-induced necrosis, 427–31
 transient global amnesia, 113, 116
- HIV infection/AIDS
 cerebrovascular disease, 196
 middle cerebellar peduncle infarction vs., 656

Index

- neurosyphilis, 169, 172
 primary lateral sclerosis vs., 40
 hoarseness, lipoid proteinosis, 268
 hot-cross-bun sign
 Machado-Joseph disease, 69, 72
 multiple system atrophy, 19–20
 HTLV-1 infection, 40, 212
 HTLV-1-related neurologic complex, 209–12
 humming bird sign, 27–8
 Hunter syndrome
 (mucopolysaccharidosis type II), 377–80
 huntingtin protein, 56
 Huntington disease (HD), 56–7, 60
 hyalinosis cutis et mucosae. *See* lipoid proteinosis
 hybrid phakomatosis, 580
 hydrocephalus
 choroid plexus papilloma, 560
 developmental venous anomalies, 128
 mucopolysaccharidosis, 380
 27-hydroxylase, mitochondrial, 52
 hyperalimentation, 84, 282
 hyperammonemic encephalopathy, acute, 301–5
 hypercoagulable state, multiple early infarcts, 110
 hyperglycemic hemiballismus and hemichorea (HHH), 84, 281–3
 hyperintense putaminal slit sign, 21–2
 hypertensive emergency, 271–2, 275
 hypertensive intracranial hemorrhage (HIC), 278
 hypertrophic olivary degeneration, 625–9
 hypoglycemia, acute, 84, 656
 hypogonadotropic hypogonadism, hereditary hemochromatosis, 345, 348
 hypomelanosis of Ito, 311–14
 hyponatremia, rapid correction, 372
 hypothalamic hamartoma
 isolated, 572
 Pallister-Hall syndrome, 570, 572
 syndromic associations, 572
 hypothalamus
 anti-Ma2-associated paraneoplastic encephalitis, 445
 Whipple disease, 18
 Wolfram syndrome, 354
 hypoxic ischemic injury (HII), 56
 idiopathic late-onset cerebellar ataxia (ILOCA), 20
 IKBKG gene, 576
 immune reconstitution
 inflammatory syndrome, progressive multifocal leukoencephalopathy-associated, 163–6
 immunosuppressed patients. *See also* HIV infection/AIDS
 aspergillosis, 186
 calcineurin inhibitor-mediated bilateral limbic injury, 599–602
 Chagas disease, 202–3
 PRES with hemorrhage, 279
 incontinentia pigmenti, 314, 576
 infantile neuroaxonal dystrophy (INAD). *See* phospholipase-associated neurodegeneration
 infarction. *See also* stroke
 developmental venous anomalies, 128
 hemorrhagic, 84, 138
 incomplete, 84–5
 isolated bilateral middle cerebellar peduncle, 653–7
 isolated corpus callosum, 308
 mammillothalamic tract degeneration after thalamic, 587–8
 secondary to hypercoagulable state, 110
 subcortical lacunar, 110
 tuberculous vasculitis, 197
 inferior olivary nuclei (ION)
 hypertrophic olivary degeneration, 628–9
 metronidazole-induced encephalopathy, 322–4
 progressive ataxia and palatal tremor, 43, 46
 internal cerebral vein (ICV)
 thrombosis, bilateral, 135–9
 intracranial hypertension, idiopathic, 593–7
 intracranial hypotension
 spontaneous (SIH), 590
 with midbrain swelling, 589–91
 intracranial pressure (ICP), 596
 inverted V appearance, subacute combined degeneration, 366
 iron deposition, 4
 aceruloplasminemia, 15–16
 BPAN, 1–2
 hereditary hemochromatosis, 348
 MPAN, 4
 multiple system atrophy, 21–2
 neuroferritinopathy, 8
 phospholipase-associated neurodegeneration, 14
 PKAN, 10
 ischemia, cerebral. *See also* infarction; small vessel ischemic disease; stroke
 bevacizumab-treated glioblastoma, 494
 ruptured dermoid cyst with, 536
 spectacular shrinking deficit, 84
 transient global amnesia, 116
 tuberculous vasculitis, 195–7
 Japanese B encephalitis, 216
 with neurocysticercosis, 215–16
 JC virus, 166
 Kayser-Fleischer (KF) rings, 342
 kinky hair syndrome. *See* Menkes disease
 Korsakoff syndrome, 116, 360
 LA2G6 gene mutations, 14
 lactate
 LBSL, 65–6
 MELAS, 336
 lamellar pattern. *See* onionskin configuration
 lateral ventricles
 enlarged, Huntington disease, 55–6
 intraventricular hemangiopericytoma, 510
 ring-shaped nodules, 615–18
 temporal horn choroid plexus papilloma, 560
 LBSL. *See* leukoencephalopathy with brainstem and spinal cord involvement and lactate elevation
 Leber hereditary optic atrophy, 354
 Leigh disease, 8, 252, 342
 leptomeningeal carcinomatosis, 152, 220
 leptomeningeal involvement
 acute cerebellitis, 155–6
 neurocysticercosis, 213–14
 neurosarcoidosis, 234
 primary angiitis of CNS, 218
 leptomeningeal melanoma, primary, 634
 leukoencephalopathy with brainstem and spinal cord involvement and lactate elevation (LBSL), 63–6, 264
 leukoencephalopathy with vanishing white matter, 261–5
 Lhermitte-Duclos disease, 404
 limbic encephalitis
 other causes, 446
 paraneoplastic, 18, 446
 limbic injury, calcineurin inhibitor-mediated bilateral, 599–602
 lingual gyrus, MELAS, 333, 336
 lipoblastic meningioma, 414
 lipoid proteinosis, 267–9
 lipoma, 514
 interhemispheric, 514
 pericallosal and left frontal surface, with cortical dysplasia, 512–15
 liponeurocytoma, cerebellar, 421–4
 liposarcoma, 414
 Listeria rhombencephalitis, 157–61, 182
 liver disease, chronic, 292
 liver, hereditary
 hemochromatosis, 347–8
 lymphoma, central nervous system (CNS)
 differential diagnosis, 90, 182, 554
 perivascular spread, 242
 lymphomatoid granulomatosis, 242
 lymphomatosis cerebri, 488
 Ma2-associated paraneoplastic encephalitis, 445–7
 Machado-Joseph disease (MJD), 72
 malaria, cerebral, 205–8
 malignant transformation
 intracranial dermoid cyst, 411–14
 neurocutaneous melanosis, 634–5
 mamillary bodies, Wernicke encephalopathy, 357, 360–1
 mammillothalamic tract
 degeneration, thalamic infarct with, 587–8
 manganese (Mn) deposition, chronic liver disease, 292
 Marchiafava-Bignami disease, 307–9
 medulla oblongata. *See also* brainstem
 adult-onset Alexander disease, 376
 hypertrophic olivary degeneration, 626, 628
 medulloblastoma, 404, 410
 cerebellar liponeurocytoma vs., 424
 molecular subgroups, 404
 multifocal desmoplastic, 407–10
 with extensive nodularity, 401–5
 megalencephalic leukoencephalopathy with subcortical cysts (MLSC), 264
 melanoblastosis cutis. *See* incontinentia pigmenti
 melanocytic nevi, congenital (CMN), 633–4

Index

- melanoma
 neurocutaneous melanosis
 transforming to, 634–5
 primary leptomeningeal, 634
- MELAS (mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes), 333–6
 differential diagnosis, 110, 252
- meningeal neurosyphilis, 172
- meningoangiomas, 383–7
- meningioma, 386, 452
 angiomatous, 452
 cavernous sinus, 530
 chondroma vs., 452, 458
 hemangiopericytoma vs., 510
 lipoblastic, 414
 microcystic, 452–3
 pineal region, 498
 sinking skin flap syndrome
 after, 603–6
 trigeminal trophic syndrome
 after, 647–50
- meningitis
 chemical, ruptured dermoid
 cyst, 536
 tuberculous, 193, 196–7, 202
- meningocele, 148
- meningovascular syphilis (MVS), 196
- Menkes disease, 295–8
- mesial temporal sclerosis, 36
- metabolic diseases, 261–5
- metachromatic leukodystrophy, 40, 264
- metastases, central nervous
 system, 480
 differential diagnosis, 182, 186
 mucinous adenocarcinoma,
 477–80
 pineal region, 498
 renal cell carcinoma in
 posterior fossa, 501–5
- methanol toxicity, 282, 342
- methotrexate encephalopathy,
 317–19
- methotrexate-induced
 leukoencephalopathy, 318
- metronidazole-induced
 encephalopathy, 46, 321–4
- MGMT (O⁶-methyl-guanine
 methyltransferase) gene
 promoter, methylation
 status, 433, 436
- Mickey Mouse appearance, 27–8
- microhemorrhages
 amyloid β -related angiitis,
 93–4
 CADASIL, 109–10
 cerebral fat embolism, 121–2
- midbrain
 atrophy, progressive
 supranuclear palsy, 27–8
 swelling, intracranial
 hypotension with, 589–91
- Wilson disease, 341–3
- middle cerebellar peduncles
 (MCPs). *See also* cerebellar
 peduncles
 fragile X-associated tremor/
 ataxia syndrome, 581, 584–5
 isolated bilateral infarcts,
 653–7
 progressive multifocal
 leukoencephalopathy, 163,
 166
 Wilson disease, 340, 342
 Wolfram syndrome, 353–4
- middle cerebral artery (MCA)
 focal stenosis, 83–4, 219–20
 temporary occlusion, 84–5
- Miller-Fisher syndrome, 240
- mineralizing microangiopathy,
 613–14
- mitochondrial diseases, 66
- mitochondrial
 encephalomyopathy with
 lactic acidosis and stroke-
 like episodes. *See* MELAS
- mitochondrial membrane
 protein-associated
 neurodegeneration
 (MPAN), 2–5
- morning glory sign, 27–8
- motor cortex, primary lateral
 sclerosis, 37, 40
- motor neuron disease, 40–1, 566,
 See also amyotrophic lateral
 sclerosis; primary lateral
 sclerosis
- motor neuropathy, MPAN, 4
- motor trephine syndrome. *See*
 sinking skin flap syndrome
- MPAN. *See* mitochondrial
 membrane protein-
 associated
 neurodegeneration
- mucinous metastasis, 182, 477–
 80
- mucopolysaccharidosis (MPS),
 377–80
- multiple sclerosis (MS)
 Balo concentric sclerosis, 246
 Marchiafava-Bignami disease
 vs., 308
 neuromyelitis optica vs., 258–9
 primary lateral sclerosis vs., 40
 Susac syndrome vs., 104
- multiple system atrophy (MSA),
 20
 cerebellar type (MSA-C), 19–20
 parkinsonism type (MSA-P),
 20–2, 56
- myoclonus epilepsy and ragged
 red fibers (MERRF), 252
- nasal ulceration, trigeminal
 trophic syndrome, 648, 650
- nasopharyngeal cancer, treated,
 427, 430
- NEMO gene, 576
- neuroacanthocytosis, 60
- neuro-Behçet disease, 227–9,
 242. *See also* Behçet disease
- neurocutaneous melanosis,
 631–5
- neurocysticercosis
 Japanese B encephalitis with,
 215–16
 racemose, 213–14
- neurocytoma, extraventricular,
 415–18
- neurodegeneration with brain
 iron accumulation (NBIA),
 4
 aceruloplasminemia, 15–16
 BPAN, 2
 MPAN (type 4), 3–5
 neuroferritinopathy, 7–8
 phospholipase-associated
 neurodegeneration, 13–14
 PKAN (type 1), 9–11
- neurodegenerative diseases, 1–2
- neuroenteric cysts, 476
- neuroferritinopathy (NFT), 7–8
 aceruloplasminemia vs., 8, 16
- neurofibromatosis type 1, with
 concomitant tuberous
 sclerosis, 580
- neuroinfectious diseases, 149–52
- neuroinflammatory diseases,
 217–21
- neuromyelitis optica spectrum
 disorder (NOSD), 255–9
- neurosarcoidosis, 234
 differential diagnosis, 182, 202,
 234, 242
 parenchymal, 231–4
- neurosyphilis
 cerebral gumma, 175–9
 differential diagnosis, 196, 446
 manifestations, 172, 178
 with temporal lobe
 involvement, 169–72
- neurovascular diseases, 78–9
- nitrous oxide (N₂O) intoxication,
 366
- non-accidental trauma (child
 abuse), 298
- NOTCH3 gene mutations, 110
- obesity, idiopathic intracranial
 hypertension, 596
- occipital lobe
 MELAS, 334–5
 microcystic meningioma, 449,
 452
- oculofacial(-skeletal)
 myorhythmia, 18
- oligodendroglioma, 90, 268, 392,
 418, 524
- olivary degeneration,
 hypertrophic, 625–9
- olivopontocerebellar atrophy,
 sporadic, 72
- onionskin configuration, Balo
 concentric sclerosis, 245–6
- optic atrophy
 MPAN, 3–4
 phospholipase-associated
 neurodegeneration, 13–14
 Wolfram syndrome, 351, 354
- optic nerve
 idiopathic intracranial
 hypertension, 593, 596
 neuromyelitis optica spectrum
 disorder, 258
- optic tracts, subacute combined
 degeneration involving,
 363–6
- osmotic demyelination syndrome
 (ODS), 272, 369–72
- Pacchionian depressions. *See*
 arachnoid granulations,
 giant
- palatal tremor
 hypertrophic olivary
 degeneration, 625, 628
 progressive ataxia and palatal
 tremor, 43, 46
- Pallister-Hall syndrome, 572
- panda sign, 343
- PANK2 gene mutations, 10
- pantothenate kinase 2 (PANK2),
 10
- pantothenate kinase-associated
 neurodegeneration
 (PKAN), 9–11
 atypical, 10
 differential diagnosis, 2, 4, 8
 typical, 10
- paradoxical herniation,
 postcraniectomy, 606
- paraneoplastic encephalitis
 syndromes
 anti-Ma2-associated
 paraneoplastic encephalitis,
 445–7
 anti-NMDA receptor
 encephalitis, 223–6
 limbic encephalitis, 18, 446
 rhombencephalitis, 160
- parental nutrition, long-term,
 282
- parietal lobe atrophy,
 corticobasal syndrome, 26
- Parkinson-plus syndrome (PPS),
 28
- Parkinson disease (PD), 2, 4,
 21–2, 28
- Parry-Romberg syndrome,
 637–41
- penguin-silhouette sign, 27–8
- peppered appearance,
 CLIPPERS, 242
- Percheron artery occlusion, 138,
 252
- periaqueductal gray, Wernicke
 encephalopathy, 358, 360

Index

- perimesencephalic subarachnoid hemorrhage (PMSAH), 117–18
- perivascular spaces, dilated
hypomelanosis of Ito, 311, 314
mucopolysaccharidosis, 377, 380
- phakomatosis, hybrid, 580
- phospholipase-associated neurodegeneration (PLAN), 13–14
atypical, 14
differential diagnosis, 2, 4
- Pick disease, 60
- pilocytic astrocytoma (PA), 548
- pilomyxoid astrocytoma (PMA), 18, 546–9
- pineal parenchymal tumors, 498
- pinealoblastoma, 498
with drop metastasis, 498–9
- pituitary gland, hereditary hemochromatosis, 346, 348
- PKAN. *See* pantothenate kinase-associated neurodegeneration
- PLAN. *See* phospholipase-associated neurodegeneration
- pleomorphic xanthoastrocytoma, 524
- polysyndactyly, Pallister-Hall syndrome, 569
- pons
central variant of PRES, 271–2
CLIPPERS, 241–2
hypertrophic olivary degeneration, 625, 628
Machado-Joseph disease, 69, 72
multiple system atrophy, 19–20
osmotic demyelination syndrome, 369, 372
Wilson disease, 341–3
Wolfram syndrome, 353–4
- posterior cortical atrophy, 26
- corticobasal syndrome, 26
- posterior fossa, renal cell carcinoma metastasis, 501–5
- posterior reversible encephalopathy syndrome (PRES), 272
atypical or severe forms, 144
calcineurin inhibitor-induced limbic injury vs., 602
central variant (CV), 271–3
hemorrhagic, 275–9
methotrexate-induced, 318
- premotor cortex, primary lateral sclerosis, 39–40
- PRES. *See* posterior reversible encephalopathy syndrome
- primary angiitis of central nervous system (PACNS), 217–21
differential diagnosis, 94, 100, 110
- primary lateral sclerosis (PLS), 40–1
- progressive ataxia and palatal tremor (PAPT)
familial, 46
idiopathic, 43–7
- progressive multifocal leukoencephalopathy (PML)
classic lesions, 166
immune reconstitution inflammatory syndrome, 163–6
middle cerebellar peduncle infarction vs., 656
- progressive nonfluent aphasia (PNFA), 60
- progressive supranuclear palsy (PSP), 28–9
classic (Richardson) variant (PSP-R), 28
corticobasal syndrome, 26
frontotemporal lobar degeneration, 60
variants, 28
- prosopagnosia, progressive (PP), 32
- protoplasmic astrocytoma, 395–9
- pseudobulbar affect, 40
- pseudotumor cerebri. *See* intracranial hypertension, idiopathic
- punica granatum seed sign, chondroma, 458
- putamen
Huntington disease, 55–7
multiple system atrophy-Parkinsonian type, 21–2
Wilson disease, 342
- pyogenic brain abscess, 186, 542, 554
- pyramidal tracts
LBSL, 64, 66
multiple system atrophy, 22
- Quattrone's magnetic resonance (MR) Parkinsonism index, 28
- rabbit ears appearance, subacute combined degeneration, 366
- radiation necrosis, delayed
hippocampus, 427–31
tumefactive cyst formation vs., 134
- radiation-induced tumefactive cyst, 131–4
- radiation-induced tumors, 134
- Ramsay Hunt syndrome, involving trigeminal spinal nucleus and tract, 149–52
- Rasmussen encephalitis, 640
- Refsum disease, 52
- renal cell carcinoma, metastasis to posterior fossa, 501–5
- retinal degeneration, 14, 16
- reversible cerebral vasoconstriction syndrome (RCVS), 97–101, 278
- reversible splenial lesion syndrome (RESLES), 327–30
- Reye syndrome, 252
- rhombencephalitis (RE). *See also* Bickerstaff brainstem encephalitis
central variant of PRES vs., 272
etiology, 160
Listeria, 157–61, 182
- right temporal lobe atrophy (RTLTA), 31–2
- right temporal variant of frontotemporal lobar degeneration, 32
- ring-shaped lateral ventricular nodules (RSLVNs), 615–18
- rosette-forming glioneuronal tumor of fourth ventricle, 381–2
- sandwich sign, 308
- sarcoidosis, nervous system. *See* neurosarcoidosis
- satellite lesions, rosette-forming glioneuronal tumor, 382
- scalp, cutis verticis gyrata, 643–6
- schistosomiasis, 189–92
- schwannoma, cavernous sinus, 530
- scleroderma, localized, 640
- Seitelberger disease. *See* phospholipase-associated neurodegeneration
- semantic dementia (SD), 32, 60
- seminoma, anti-Ma2-associated
paraneoplastic encephalitis, 445–7
- sinking skin flap syndrome, 603–6
- sinus pericranii, 145–8
- skeletal manifestations
Erdheim-Chester disease, 621–2
Menkes disease, 298
mucopolysaccharidosis, 379–80
Pallister-Hall syndrome, 569
- skin manifestations
hypomelanosis of Ito, 313–14
incontinentia pigmenti, 573, 575–6
lipoid proteinosis, 267–8
neurocutaneous melanosis, 633–4
- small vessel ischemic disease (SVID), 318, 488
- snowball-like lesions, Susac syndrome, 103–5
- spastic paraplegia, hereditary. *See* hereditary spastic paraplegia
- spectacular shrinking deficit (SSD), 84–5
- SPG gene mutations, 566
- spinal cord. *See also* cervical spinal cord
HTLV-1-related tropical spastic paraparesis, 212
LBSL, 65–6
neuromyelitis optica spectrum disorder, 257–9
subacute combined degeneration, 363–6
- spine
drop metastases, 496, 498, 503–4
mucopolysaccharidosis, 379–80
- spinocerebellar ataxia (SCA)
type-3. *See* Machado-Joseph disease
type-6, 72
- splenium of corpus callosum
metronidazole-induced encephalopathy, 321, 323–4
reversible/transient lesion, 327–30
- sporadic subcortical arteriosclerotic encephalopathy (SSAE), 110
- squamous cell cancer, dermoid cyst transformation to, 414
- star field pattern, cerebral fat embolism, 122
- static encephalopathy (of childhood) with neurodegeneration in adulthood (SENDA). *See* beta-propeller protein-associated neurodegeneration
- striatum
atrophy, Huntington disease, 56–7
hyperglycemic hemiballismus and hemichorea, 281–2
- string-of-beads appearance, reversible cerebral vasoconstriction syndrome, 101
- string-of-pearls appearance, Susac syndrome, 103–5
- stroke. *See also* infarction; ischemia, cerebral
CADASIL, 110
MELAS, 336
racemose neurocysticercosis, 213–14
reversible cerebral vasoconstriction syndrome, 100–1
ruptured dermoid cyst, 535–6
tuberculous vasculitis, 197
- subacute combined degeneration, with optic tract involvement, 363–6
- subarachnoid hemorrhage (SAH) aneurysmal, 100

Index

- subarachnoid hemorrhage (SAH) (cont.)
 - perimesencephalic (PMSAH), 117–18
 - PRES with, 277–8
 - reversible cerebral vasoconstriction syndrome, 97, 100
- subarachnoid spaces
 - enhancing lesions, 202
 - enlarged, 55–6
- subcortical arteriosclerotic encephalopathy, sporadic (SSAE), 110
- subdural collections
 - Menkes disease, 295, 298
 - spontaneous intracranial hypotension, 590
- subependymal heterotopia, 618
- subependymal nodules, 618
- subependymoma, 90, 560, 618
- substantia nigra
 - aceruloplasminemia, 15–16
 - iron deposition, 1–2
 - neuroferritinopathy, 7
 - PKAN, 10
- superior cerebellar artery (SCA), 656
- Susac syndrome (SS), 103–5, 110, 308
- Swiss cheese pattern, radiation necrosis, 429–30
- syphilis, 172, 178, 196, *See also* neurosyphilis
 - cerebral gumma, 175–9
- T2-dark through effect, white epidermoid cyst, 476
- tabes dorsalis, 172
- tacrolimus-mediated bilateral limbic injury, 599–602
- target sign, tuberculoma, 554
- tauopathies, 26, 28
- temporal horn choroid plexus papilloma, 560
- temporal lobe
 - atrophy, right-sided (RTLA), 31–2
 - neurosyphilis, 169–72
 - radiation-induced necrosis, 427–31
 - Whipple disease, 18
- testicular tumor, anti-Ma2-associated paraneoplastic encephalitis, 445–7
- thalamus
 - aceruloplasminemia, 15–16
 - anti-Ma2-associated paraneoplastic encephalitis, 445
 - infarct, with
 - mammillothalamic tract degeneration, 587–8
 - internal cerebral vein thrombosis, 136, 138
 - Wernicke encephalopathy, 358, 360
 - Whipple disease, 18
- thiamine deficiency, 360
- third ventricle
 - craniopharyngioma, 461–5
- TORCH group of infections, 314
- toxoplasmosis, 202
- transient global amnesia (TGA), 113–16
- transient ischemic attacks (TIA), multiple early, 107, 110
- transient splenial lesion, 327–30
- trichothiodystrophy. *See* Menkes disease
- trigeminal nerve (CN V)
 - pathways
 - LBSL, 66
 - Ramsay Hunt syndrome involving, 149–52
- trigeminal trophic syndrome (TTS), 647–50
- tropical spastic paraparesis, 209–12
- tuberculoma, giant, 554–5
- tuberculosis, 196–7
 - cerebral vasculitis, 193–7
 - meningitis, 193, 196–7, 202
- tuberous sclerosis
 - hypomelanosis of Ito vs., 314
 - subependymal nodules, 618
 - with concomitant neurofibromatosis type 1, 580
- tumefactive cyst, radiation-induced, 131–4
- tumefactive demyelination, 272
- tumors, central nervous system, 381–2
 - Balo concentric sclerosis vs., 246
 - calcification, 90, 268
 - fat-containing, 414, 424
 - radiation-induced, 134
 - syphilitic gumma resembling, 178–9
- ulceration en arc, 650
- Urbach Wiethe disease. *See* lipid proteinosis
- vanishing white matter (VWM) disease, 261–5
- varicella-zoster virus (VZV), 152, 156
- vascular dementia (VD), 36
- vasculitis, cerebral. *See also* primary angiitis of central nervous system
 - neurocysticercosis, 214
 - primary, 228
 - tuberculosis, 193–7
- vasogenic edema. *See also* edema, cerebral
 - cerebral amyloidoma, 87–90
 - CNS aspergillosis, 183, 186
 - PRES with hemorrhage, 275, 277–8
- venous malformations. *See also* sinus pericranii
 - complications, 125–9
 - hypertrophic olivary degeneration, 625, 628
 - meningoangiomatosis vs., 386
- venous sinuses, dural. *See* dural venous sinuses
- venous thrombosis, cerebral. *See* cerebral venous thrombosis
- viral encephalitis. *See also* Japanese B encephalitis
 - differential diagnosis, 104, 160, 488
 - hemorrhagic, 208
- visual disturbances. *See also* optic atrophy
 - idiopathic intracranial hypertension, 593, 596
 - MELAS, 333
 - subacute combined degeneration, 366
 - Susac syndrome, 105
- vitamin B12 deficiency, 40, 66, 366
- Wernicke encephalopathy, 324, 357–61
- WFS1 (wolframin) gene, 354
- Whipple disease (WD), 17–18
- whirlpool-like pattern, epidermoid cyst, 470
- white epidermoid cyst, 473–6
- Wilson disease (WD), 8, 339–43, 584
- wine glass appearance, 40
- Wolfram syndrome, 351–4
- xanthoastrocytoma, pleomorphic, 524