ACR APPROPRIATENESS CRITERIA

Ataxia

J.A. Brunberg,
for the Expert Panel on
Neurologic Imaging

Ataxia is manifested by a wide-based unsteady gait, errors of extremity trajectory or placement, errors in motor sequence or rhythm and/or by dysarthria.[1] Tone is usually decreased and stretch reflexes may be "pendular." Nystagmus, skew deviation, disconjugate saccades, and altered ocular pursuit can be present. Truncal instability and tremor of the body or head may occur, especially with cerebellar midline disorders.[1]

Ataxia can arise from disorders that involve cerebellum, spinal cord, brain stem, vestibular nuclei, thalamic nuclei, cerebral white matter, cortex (especially frontal), and peripheral sensory nerves. Because symptoms and signs are difficult to localize, imaging may be extensive and complex.

Disorders causing ataxia are numerous and often uncommon and recommendations for imaging are imprecise. The purpose here is to categorize the disorders that present with ataxia, and to suggest imaging objectives. Clinical and family history, physical findings and laboratory studies are essential.

For basic imaging purposes ataxia is approached on the basis of symptoms, age at onset, potential disease mechanism, and urgency for excluding disorders that requires immediate management. The ACR Appropriateness Criteria on Ataxia is summarized below.

Classification of Disorders Causing Ataxia

Mass Lesions

In pediatric patients the most common posterior fossa lesions are medulloblastoma, cystic astrocytoma, ependymoma, and brain stem glioma. In adults; hemangioblastomas, choroid plexus papillomas, extra-axial meningiomas, and metastatic processes become more prevalent. Frontal lobe and thalamic mass lesions can present with ataxia. MR imaging, without and with contrast, is superior to CT for these disorders.

Lhermitte-Duclos disease (dysplastic gangliocytoma) is a benign cerebellar hamartoma .[2] Symptoms relate to local mass effect. MR imaging demonstrates a nonenhancing mass involving cortex and folia, with increased T2 signal intensity (SI) and internal curvilinear lower intensity bands.[2] The mass may demonstrate restricted diffusion.[2]

Paraneoplastic cerebellar degeneration is characterized by acute or subacute ataxia, dysarthria, and ocular dysmetria.[3] It usually occurs with breast, gynecologic, and lung tumors, or with Hodgkin disease.[3] Antineuronal antibodies are identified in serum. MR imaging is generally normal until late, when cerebellar cortical atrophy becomes evident.[3] Uncommonly, there is increased T2 SI in cerebellar or other areas. Evaluation for an underlying primary is necessary.

Vascular Disorders

Hemorrhage or infarction localized to the cerebellum, lateral medulla or pons, mesencephalon, red nucleus, thalamic nuclei, posterior limb of the internal capsule, or to frontal or parietal cortex can result in ataxia.[4] Syndromes associated with infarction[5] in the posterior inferior cerebellar artery territory (lateral medullary or Wallenberg syndrome) include ipsilateral hemiataxia, vertigo, dysarthria, ptosis, and miosis. Evaluation generally requires MR imaging, diffusion characterization and MR angiography. Neck vessel MR imaging may exclude dissection.[6] MR venography helps when central or dural venous thrombosis is considered. Catheter-based and/or CT angiography (CTA) is occasionally necessary.

In superficial siderosis hemosiderin accumulates in subpial brain and spinal cord as the result of recurrent, often silent, subarachnoid hemorrhage. Symptoms include slowly progressive ataxia and hearing loss.[7] MR imaging shows low superficial T2 SI over cortex, brain stem, and/or spinal cord with usual cerebellar atrophy.

Infectious and Postinfectious Processes

Bacterial cerebellitis can occur with meningitis, penetrating trauma or extension of an epidural process, most commonly from temporal bone. Diffusion imaging and MR spectroscopy narrow the differential diagnosis.[8] Viral processes, including herpes and arborvirus, can also involve brain stem or cerebellum.[9]

Prion-associated encephalopathies include sporadic Creutzfeldt-Jakob disease (sCJD), variant Creutzfeldt-Jakob disease (vCJD) or bovine spongiform encephalopathy, and familial Creutzfeldt-Jakob disease (fCJD). sCJD and vCJD present with behavioral, emotional, and intellectual deterioration, followed by ataxia and dysarthria. Progression is to stupor and coma. MR imaging demonstrates increased T2 SI and diffusion-weighted SI in the heads of the caudate nuclei, the putamen, and in regions of frontal, parietal, and occipital cortex. Findings can initially be asymmetric.[10] There is eventual diffuse volume loss. While all forms of CJD can have increased T2 SI and restricted diffusion in the thalamic nuclei and pulvinar, this alteration is prominent in vCJD.[10]

Acute cerebellitis (acute cerebellar ataxia) is a para-infectious disorder, predominately of childhood. Symptoms include headache, ataxia, photophobia, and increased intracranial pressure. MR imaging demonstrates mass effect and increased cerebellar T2 SI.[11] Lateral ventricle enlargement, up-

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Please address correspondence to James A. Brunberg, MD, Department of Radiology, University of California-Davis Medical Center, 4860 Y St, ACC Bldg, Suite 3100, Sacramento, CA 95817-2309; e-mail: brunberg@ucdavis.edu

Clinical condition: ataxia									
Duration of ataxia (adult or child)	MRI head	MRI cervical, thoracic and lumbar spine	MRI cervical spine	CT head	CT head without contrast	MRA head	MRA head and neck	MR spectroscopy head (MRS)	CTA head and/or neck
Slowly progressive or chronic*	8	7[a]	X	5	X	X	X	2	X
Acute (< 3 hours) as a suspected stroke†	8[b,c]	X	5[g]	8[b,d]	X	X	8[b]	2	8[b]
Acute or subacute as a suspected infection	8	X	6[a]	5[e]	4	5	X	6[f]	5
Acute following head trauma‡	8	X	X	6[e]	9	X	6	X	6

Note:—Rating Scale: 1, least appropriate; 9, most appropriate.
* FDG-PET brain received a rating of 3.
† See the ACR Appropriateness Criteria topic for cerebrovascular disease.
‡ MRI neck received a rating of 6.
[a] Ataxia can be of spinal origin. Consider if brain imaging is negative or inconclusive.
[b] MR preferred if treatment is not unreasonably delayed. Combined vascular and cerebral evaluation should be considered.
[c] Fat saturated T1 axial images.
[d] CT perfusion is less accurate in the posterior fossa.
[e] CT temporal bone may be useful when skull-based or middle ear disease suspected.
[f] May help distinguish abscess from other masses.

ward herniation and meningeal enhancement may occur. Follow-up imaging may demonstrate cerebellar atrophy.[12]

Bickerstaff encephalitis is a postviral brain stem and cerebellar inflammation with ataxia and ophthalmoplegia. MR imaging demonstrates brain stem and cerebellar mass effect with increased T2 SI.[13]

Fisher syndrome, a Guillain-Barré variant, involves the peripheral and central nervous system. Ophthalmoplegia and ataxia are associated with transient high T2 SI in the cerebellum and/or brain stem.[14] Cranial nerves and spinal nerve root enhancement can be seen with increased T2 SI in posterior spinal cord.[15] There is late cerebellar atrophy.

Trauma

Gait instability may persist following cerebellar, vestibular, brain stem, or frontal lobe injury or with interruption of the frontopontocerebellar tract.[16] In acute trauma, or progressive post-traumatic ataxia, an expanding cyst or extra-axial hematoma should be considered.

Demyelinating Disorders

Ataxia is common in early and late multiple sclerosis. MR findings have been reviewed.[17]

Congenital Disorders

Dandy Walker Syndrome, with ataxia, nystagmus, cranial nerve palsies, apneic episodes, hydrocephalus, and cognitive dysfunction, demonstrates hypoplasia of the cerebellar vermis and a CSF collection that is predominately posterior to the cerebellum but continuous with the fourth ventricle.[18] The torcula is usually elevated and the posterior fossa enlarged. Hydrocephalus is common, and anomalies of cerebral development may involve cortex and corpus callosum.[18] Differentiation from congenital or acquired posterior fossa cysts is essential.

Joubert syndrome, with congenital ataxia, hypotonia, and oculomotor ataxia, has unique imaging alterations that include a "molar tooth" contour of brain stem and "bat wing" configuration of the fourth ventricle.[19] There are 4 sub-types with variable clinical and imaging features, and with differing genetic alterations.[20]

Rhombencephalosynapsis is a rare cerebellar dysplastic process that can occur alone or in association with other developmental anomalies. There is vermian agenesis, fusion of the cerebellar hemispheres, and fusion of the superior cerebellar peduncles. Lateral ventricles are usually enlarged, and there may be fusion of the thalamic nuclei. Symptoms are variable.

Ataxia can also occur with perinatal cerebral infarction and with congenital CNS infection.[18,21] Ataxic cerebral palsy is uncommon and imaging findings are variable.[22]

Hereditary and Idiopathic Degenerative Processes

Hereditary ataxias are classified by the causative gene (when known) and their pattern of inheritance. MR imaging is preferred. A broad range of diagnoses is often suggested by family history, by findings on physical examination, and MR evidence of atrophy involving cerebellum and varying combinations of the pons, medulla, spinal cord, cerebral cortex and optic nerves. Dentate calcification may be identified. Definitive diagnosis relies on molecular genetic testing. While ataxia is the identifying clinical finding, spasticity, neuropathy, seizures, extrapyramidal symptomatology, cognitive decline, nystagmus, visual loss, spasmodic cough, and migrainelike episodes may be associated.

Among autosomal dominant spinocerebellar ataxias (AD-SCAs), specific diagnostic nomenclature is replacing terms like "spinocerebellar degeneration," "Marie's ataxia," and OPCA. Among AD-SCA disorders, 22 genetic abnormalities have been identified. "OPCA" is now used only for cases with a combination of "cerebellar-plus" symptomatology, imaging correlates of cerebellar and brain stem atrophy, and an unidentified genetic basis.[23] "Idiopathic late onset cerebellar ataxia" is used to describe a different large group of adults with predominant cerebellar symptomatology, absence of a family history, and absence of an identified genetic marker.[24] Imaging generally demonstrates cerebellar and pontine volume loss.

Other genetic and sporadic neurodegenerative disorders need to be considered in the evaluation of slowly progressive ataxia. AD SCAs also include SCA2, SCA3 (Machado-Joseph disease), and Dentorubral-pallidolusian atrophy (DRPLA).[25] Autosomal recessive disorders associated with ataxia include Friedreich's ataxia and Ataxia-telangiectasia (A-T).[26-28] An x-linked disorder, Fragile X tremor/ataxia syndrome (FXTAS), has recently been described.[29-31] Multiple system atrophy (MSA) is a sporadic disorder that initially manifests after age 50 by ataxia (MSAc) or Parkinsonism (MSAp).[29,32] Mitochondrial disorders are also associated with progressive ataxia, with coenzyme Q deficiency and an early childhood leukoencephalopathy known as vanishing white matter disease being examples. Clinical and imaging findings on these disorders are further characterized in the ACR Appropriateness Criteria on Ataxia (www.acr.org/ac).

Paroxysmal Disorders Associated with Ataxia

Intermittent ataxia has been associated with epilepsy, migraine, and high systemic fever in otherwise healthy children. Intermittent ataxia can also be associated with abnormalities in membrane calcium or potassium channel function, or with altered synaptic glutamate transport.[33,34] MR imaging may be normal, may demonstrate cerebellar volume loss, or may demonstrate areas of cortical increased T2 SI that may correlate with the possible simultaneous occurrence of hemiplegic migraine or recent seizure activity. MR imaging is the technique of choice.

Spinal Cord and Peripheral Nerve-Related Ataxia

Spinal cord and/or nerve root disorders can produce ataxia. High resolution T1 and T2 axial and sagittal imaging, without and with contrast needs to focus on posterior columns and on nerve roots. In pernicious anemia there may be early localized or relatively diffuse cord swelling with increased T2 SI that is most evident in the posterior columns.[35] Late atrophy and persistent gliosis may develop, or all findings may resolve with treatment.[35] In the presence of hypertrophic, inflammatory, or postinfectious polyneuropathies, nerve root enhancement and enlargement may be demonstrated with MR imaging.[17]

Nutritional Deficiency, Toxins and Drugs

Solvent abuse or toxic exposure to solvents, methyl-mercury poisoning (Minamata disease), metronidazole (Flagyl)-induced cerebellar toxicity, central pontine myelinolysis (osmotic demyelination syndrome), leukoencephalopathy relating to the inhalation of heroin vapors, vitamin E deficiency, chronic ethanol abuse, Wernicke encephalopathy, and reversible posterior leukoencephalopathy can each be associated with the acute or chronic presentation of ataxia. In each of these disorders MR imaging is preferred. These disorders and their imaging are further characterized in the published ACR Appropriateness Criteria on Ataxia (www.acr.org/ac).

Appendix

Expert Panel on Neurologic Imaging: James A. Brunberg, MD, Principal Author, University of California-Davis Medical Center, Sacramento, Calif; David J. Seidenwurm, MD, Panel Chair; Patricia C. Davis, MD; Robert L. DeLaPaz, MD; Pr. Didier Dormont; David B. Hackney, MD; John E. Jordan, MD; John P. Karis, MD; Suresh Kumar Mukherji, MD; Patrick A. Turski, MD; Franz J. Wippold II, MD; Robert D. Zimmerman, MD; Michael W. McDermott, MD, American Association of Neurological Surgeons; Michael A. Sloan, MD, MS, American Academy of Neurology.

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