Imaging Ataxia: How, What and Where to look?

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Learning objectives

Ataxia is an abnormality in the coordination of movement associated to disorders in a large number of anatomic regions. Knowledge of the anatomy of regions related to ataxia, and the use of an appropriate algorithmic approach for radiological evaluation is essential for diagnosis. The purposes of this exhibit are:

1. Describe the different types of ataxia
2. Illustrate neuroanatomy of the motor coordination ways
3. Review different causes of ataxia and their imaging appearances.

Background

Ataxia definition

The term ataxia refers to poorly coordinated movements. It is a disorder of motor coordination that affects the amplitude and direction of movements with altered gait and associated disequilibrium.

Usually results from lesions of the cerebellum and/or cerebellar connections, but in addition to cerebellar ataxia there are also cases of sensory or vestibular ataxia caused by lesions of spinal proprioceptive pathways or the vestibular system, respectively.

The different types of ataxia have specific clinical features (table 1), depending on the nature and location of the underlying lesion:

Types of Ataxia:

- Cerebellar Ataxia:

Cerebellar ataxia manifestations commonly include a wide-based unsteady gait as well as poor coordination or clumsy movements of the extremities (dyssynergy).

Patients stand with their legs farther apart than normal and sway or fall in attempts to stand with the feet together. Mild gait ataxia may be exaggerated when the patient attempt tandem - walking in a straight line.
Cerebellar ataxia is frequently associated to slowness of movements and dysdiadochokinesia (impaired ability to perform rapid, alternating movements). Abnormal motor trajectory or placement during active movement (dysmetria with hypometria or hypermetria) and muscle hypotonia (decreased resistance to passive movement) are frequent. Muscle reflexes usually have a pendular pattern.

Involvement of the dentate nucleus or its efferent fibers causes intention tremor with increasing amplitude of the oscillations when is approaching the target.

There can be poor coordination of speech presenting as dysarthria (slurred speech or scanning speech, sometimes characterized by explosive variations in voice intensity despite a regular rhythm) and abnormality in ocular motility presenting as saccadic ocular dysmetria and horizontal nystagmus.

Relatively isolated trunk ataxia with abnormal stance and gait and axial or head tremor (titubation) is usually associated to disorders that involve the cerebellar midline or vermis.

Extremities ataxia is usually seen in disorders that affect the cerebellar hemispheres. With unilateral cerebellar hemisphere lesions an ipsilateral disturbance of gait, posture and movement are observed.

- **Sensory Ataxia:**

Sensory ataxia is less frequent than cerebellar ataxia and is due to loss of proprioception. It could be central (due to lesions of the somatosensory cortex, the thalamocortical pathways, the thalamus, and medial lemniscus pathway), from disorders of the posterior columns of medulla (lesions of the afferent somatosensory pathways in the dorsal portion of the spinal cord - fasciculus gracilis and fasciculus cuneatus) or peripheral (caused by disease processes affecting the peripheral sensory nerves).

Clinically, it manifests with proprioceptive deficit (loss of position sense and inability to detect vibrations), an unsteady "stomping" gait, and loss of balance and significant worsening of clumsiness with eye closure. There is a positive Romberg's sign (loss of balance in a patient standing up when close the eyes). The Romberg's sign is caused because the patient can still maintain balance by using vision and vestibular function. Vision is able to compensate for the loss of position sense to a great degree and thus minimizes sensory ataxia.

Additionally, a phenomenon of "pseudoathetosis" can be seen in affected extremities in sensory ataxia. Pseudoathetosis is an abnormal writhing movements caused by a failure of joints position sense or proprioception, usually observed in fingers. Analogous to Romberg's sign, pseudoathetosis is most pronounced with eye closure.

- **Vestibular Ataxia:**
Vestibular ataxia is rare and may be classified as a specific subtype of sensory ataxia. In vestibular ataxia there are serious stance and gait difficulties (vestibular disequilibrium), but there aren't alterations in coordination of extremities or in speech. With a unilateral vestibular lesion a "flank walking" in the direction of the altered side is observed. Additionally, vestibular ataxia could be associated with nystagmus and vertigo with prominent dizziness, nausea and vomiting, particularly in acute cases. Sometimes vestibular ataxia is associated with hearing loss.

Anatomic Review:

The smooth and precise execution of a movement requires a properly functioning "regulatory system". The regulatory system (cerebellum) must integrate proprioceptive input from conscious and unconscious proprioceptive pathways, along with further input from the vestibular and visual systems, and the use of this data to optimize and coordinate each phase of the movement and plan its force and amplitude.

- Proprioceptive System:

It includes the pathways responsible for carrying sensorial proprioceptive information to the parietal cortex (conscious proprioceptive pathway) and cerebellum (unconscious proprioceptive pathway).

1. Conscious Proprioceptive Pathway: (Fig. 1, 2 and 3)

This pathway carries information from exteroceptors (tactile sense, stereognosis and vibration) and proprioceptors (position sense) through peripheral nerves and the dorsal root of spinal nerves (spinal ganglion cells - first sensory neuron). Then it travels by way of the posterior columns of spinal cord to the nucleus gracilis and nucleus cuneatus of the medulla, without any intervening relay in the spinal cord. These medullary nuclei contains the second sensory neuron, whose axons decussate to the contralateral side of the stem brain and ascend in the medial lemniscus, which travels to the ventral posterolateral nucleus of the thalamus. From there, the third - order neurons project to the somatosensory (parietal) cortex.

2. Unconscious Proprioceptive Pathway: (Fig. 4 and 5)

Unconscious proprioceptive pathway includes four fiber tracts that run up the spinal cord to the cerebellum. These are: 1. the ventral spinocerebellar tract, 2. the dorsal spinocerebellar tract, 3. the rostral spinocerebellar tract, and 4. the cuneocerebellar tract.
The dorsal and ventral spinocerebellar tracts run up the spinal cord near the dorsolateral and ventrolateral surfaces respectively. Both tracts terminate in the vermis. The dorsal spinocerebellar tract rises ipsilaterally and enters the cerebellum through the inferior cerebellar peduncle. The ventral spinocerebellar tract decussates to the contralateral ventral cord, and ascends to the cerebellum entering through the superior cerebellar peduncle.

The rostral spinocerebellar tract arises from the cervical portion of the cord, rises ipsilaterally and enters the cerebellum through the superior and the inferior cerebellar peduncles.

The cuneocerebellar tract reaches the accessory cuneate nucleus (immediately above the cuneate nucleus) through the cuneate fasciculus and synapse with the second-order fibers of the cuneocerebellar tract. These fibers ascend to the cerebellum via the inferior cerebellar peduncle.

- **Vestibular System:** (Fig. 6)

The vestibular receptors are located in the semicircular canals, utricle and saccule of the inner ear. They are mechanoreceptors that detect changes in the motion and position of the head. This information is transmitted by cranial nerve VIII to the vestibular nuclei (in the dorsal pontomedullary junction).

- **Cerebellum:**

Cerebellum participates in optimizing the amplitude, speed, and precision of voluntary movement and simultaneously in regulating the motor control of balance and adapt muscle tone. It also plays a role in the regulation of gaze-related movements of the eyes and in ensuring the smooth complementary functioning of agonist and antagonist muscle groups.

To perform these coordinating tasks, the cerebellum requires information from different parts of the nervous system:

- **Cerebral cortex (sensorial and motor cortical areas)** information for the initiation and planning of voluntary movement travels in the corticopontocerebellar pathway, by way of the middle cerebellar peduncle, to the Neocerebellum or Pontocerebellum (located in the cerebellar hemispheres and dentate nucleus). (Fig. 7)

The lateral cerebellar cortex receives its major inputs from the contralateral pontine nuclei through the middle cerebellar peduncle. The pontine nuclei receive inputs from the ipsilateral cerebral cortex. Another important afferent pathway to the neocerebellum is the olivocerebellar pathway from the contralateral principal olivary nucleus.
The major outputs of the pontocerebellum are, corticonuclear fibers from the hemisphere cerebellar cortex to the dentate nucleus and from this to the contralateral red nucleus (dentatorubral fibers), and to the VL thalamus (dentatothalamic fibers). From the VL nucleus, fibers project extensively to numerous regions of the cerebral pre-motor and motor cortex. Dentate nucleus also sends fibers to the contralateral principal olivary nucleus (dentato-olivary fibers) and, in lesser extent, to contralateral pontine and reticular formation nuclei.

This part of the cerebellum is mainly responsible for the fine control - very precise movements, particularly of the limbs and of the motor apparatus of speech.

Injury to the pontocerebellum (cerebellar hemispheres) can result in severe disruption of movement. The ipsilateral side is affected because pontocerebellar efferents project to the contralateral motor cortex, which in turn projects caudally in corticospinal fibers that decussate in the pyramids.

**- Information regarding joint position and muscle tone** from peripheral proprioceptors travels, by way of the ventral and dorsal spinocerebellar tracts and the cuneocerebellar tract, through the inferior and superior cerebellar peduncles to the **Paleocerebellum or Spinocerebellum** (superior vermis, part of inferior vermis and Globose and Emboliform nuclei). The spinocerebellum integrates the unconscious proprioceptive information and projects it to the cerebral cortex via the VL nucleus of the thalamus. There are also efferent connections to the reticular formation, vestibular nuclei and red nucleus (cerebellorubral fibers). This part of the cerebellum is mainly responsible for the smooth and synergistic functioning of the muscles when the individual stands or walks and of postural tone. *(Fig. 4 and 5)*

**- Impulses from the vestibular system** travel by way of the inferior cerebellar peduncle to the **Archicerebellum or Vestibulocerebellum** (nodulus, flocculus, adjacent vermian cerebellar cortex and the fastigial nucleus). *(Fig. 7)*

This part of cerebellum also receives impulses from visual system (lateral geniculate body). The vestibulocerebellum sends efferent connections to the fastigial nucleus and from this to the vestibular nuclei and reticular formation. From the vestibular nuclei fibers descend as the medial and lateral vestibulospinal tracts. The vermis projects also to the contralateral VL thalamus, and from there to the trunk areas of the motor cortex. This part of the cerebellum is implied in keep balance during standing and walking.

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**Table 1:** Types of Ataxia
Fig. 1: Conscious Proprioceptive Pathway
Fig. 2: Conscious Proprioceptive Pathway
**Fig. 3:** Conscious Proprioceptive Pathway
Fig. 4: Unconscious Proprioceptive Pathway - Spinocerebellum
Fig. 5: Unconscious Proprioceptive Pathway - Spinocerebellum
Fig. 6: Vestibular System

Vestibular System

Inner ear (semicircular canals, utricle, and saccule) - cranial nerve VIII - brain stem vestibular nuclei.

Lateral semicircular canal

Vestibule (Utricle and Saccule)

Vestibulocochlear nerve (CN8)
Fig. 7: Pontocerebellum and Vestibulocerebellum
Imaging findings OR Procedure details

Classification of Disorders Causing Ataxia in Adults

Cerebellar Ataxia:

Cerebellar ataxia is the result of disorder affecting the cerebellum, or its afferent or efferent tracts.

- Long Duration or Gradually Progressive Cerebellar Ataxia:

Chronic ataxia is the largest group and includes ataxia associated with intracranial mass lesions, paraneoplastic syndromes, congenital and hereditary disorders, multiple sclerosis, idiopathic degenerative processes, nutritional deficiencies, toxins and drugs.

  - Mass lesions: (Fig 8 - 12)

The exclusion of posterior fossa masses is usually one of the first steps in chronic cerebellar ataxia evaluation. In adults, the most common posterior cranial fossa masses associated to cerebellar ataxia (affecting cerebellar parenchyma or brainstem) are metastases, hemangioblastomas, medulloblastomas, and infiltrating astrocytic tumor of brainstem (Astrocytoma and Glioblastoma). Large extra axial masses as meningiomas or epidermoid cysts can be a cause of ataxia by compressive effect in cerebellar or brainstem parenchyma. Frontal lobe and thalamic masses can also present with cerebellar ataxia.

  - Paraneoplastic Cerebellar Degeneration

Paraneoplastic cerebellar degeneration usually presents with subacute ataxia. It can occur with any tumor but is most frequent in breast, gynecologic and lung tumors and in association with Hodgkin's disease. It is caused by antineuronal antibodies.

  - Congenital Disorders: (Fig. 13)

The most common congenital disorders that can cause ataxia include Dandy Walker syndrome, Joubert syndrome and Rhombencephalosynapsis. These disorders usually manifest during early childhood, but these patients can reach adult age.

  - Hereditary Disorders associated to ataxia:

Although genetic testing is mandatory for hereditary ataxias; family history, physical examination and MRI findings help in the diagnosis.
Hereditary ataxias are a clinically and genetically heterogeneous group of disorders transmitted as autosomal dominant or autosomal recessive traits. Autosomal dominant spinocerebellar ataxias (AD - SCA) are the largest group of hereditary disorders associated to ataxia. Spinocerebellar ataxias involve not only the cerebellum, but also the spinocerebellar tracts of the spinal cord. (Fig. 14)

Other hereditary ataxias include dentorubral-pallidoluysian atrophy and autosomal recessive spinocerebellar ataxias as Friedreich's ataxia (which causes a "mixed" sensory/cerebellar ataxia with predominance of sensory ataxia) and ataxia - telangiectasia syndrome (also causes a "mixed" sensory/cerebellar ataxia but with predominance of the latter).

In Friedreich's ataxia the major pathological findings are cell loss in the dentate nucleus, and combined degeneration of the spinocerebellar tracts, pyramidal tracts and posterior columns. The disease usually becomes clinically presents in the second decade of life; initially with signs of posterior column degeneration followed by spasticity and cerebellar signs. Typical findings on physical examination include, progressive sensorial ataxia with impaired proprioception; and in advanced stages of the disease, cerebellar ataxia and dysarthria. (Fig.15)

- **Demyelinating Disorders:**

In adults, this group is almost exclusively represented by multiple sclerosis. It typically presents as chronic or subacute cerebellar ataxia, especially at a young age. A relapsing-remitting course and the presence of multiple demyelinating lesions in the brain and/or spine on MRI, suggest the diagnosis. Multiple sclerosis may cause cerebellar ataxia when it affects the cerebellum, or its afferent or efferent pathways, including supratentorial, brainstem or spinal cord levels or causes sensorial ataxia when it affects the conscious proprioceptive pathway. (Fig. 16)

- **Degenerative Processes:** this group includes:

  - "Idiopathic late - onset cerebellar ataxia" or ILOCA, which is a disorder that affects adult patients and produces predominantly, cerebellar symptomatology, in the absence of family history and an identified genetic marker.

  - Multiple System Atrophy (MSA), which is a sporadic progressive neurodegenerative disorder of adult onset, that usually presents after the fifth decade of life as a combination of cerebellar, pyramidal, extrapyramidal and autonomic alterations. Parkinsonian features predominate in 80% of patients (MSA-P subtype) whereas cerebellar symptoms, with gait and limb ataxia, dysarthria and cerebellar oculomotor disturbance, predominate in 20% of patients (MSA-C subtype). Autonomic symptoms and cognitive dysfunction are frequent. (Fig. 17)
- Subcortical arteriosclerotic encephalopathy, which is associated with hypertension, and shows diffuse and confluent regions of periventricular white matter involvement (leukoariosis).

  - **Nutritional Deficiency, Toxins and Drugs:**

    This group includes exposure to toxins, as solvents and methylmercury; and to different drugs, especially some anxiolytic and anticonvulsant medications. *(Fig. 18)*

    Central pontine myelinolisis can be a cause of ataxia in chronic alcoholics or malnourished patients following rapid correction of hyponatremia; although, it most frequently presents with coma, locked-in syndrome or quadriplegia. *(Fig. 19)*

    Chronic alcoholism can cause ataxia in three different ways: by neurotoxicity of ethanol and its metabolic products *(Fig. 20)*, by associated chronic liver disease, or by thiamine deficiency (Wernicke encephalopathy).

  - **Late Postsurgical or Posttraumatic changes:**

    Gliosis and malacic changes in cerebellar as well as in afferent and efferent cerebellar tracts can be a cause of long duration ataxia. *(Fig. 21)*

- **Acute Cerebellar Ataxia as a Possible Manifestation of Stroke:** *(Fig. 22 and 23)*

  Hemorrhage or infarction resulting in ataxia can occur in the cerebellum, lateral portion of the medulla or pons, mesencephalon, red or thalamic nuclei, posterior limb of the internal capsule, or frontal or parietal cortex. Wallenberg or lateral medullary syndrome is secondary to infarction in the posterior inferior cerebellar artery (PICA) territory, and is associated with ipsilateral hemiataxia.

  Ischemic causes include lacunar, cardioembolic, or atherothrombotic infarcts, and venous thrombosis. Hemorrhagic causes that should be considered are hypertensive hemorrhagic stroke in the first place, but also vascular malformations and venous thrombosis.

- **Acute Cerebellar Ataxia as a Possible Manifestation of Infection:** *(Fig. 24 and 25)*

  Among the most important infectious causes of ataxia are bacterial cerebellitis, cerebellar abscess and prion-associated encephalopathies as Creutzfeldt-Jakob disease. Among parainfectious disorders are acute cerebellitis, which is more common in children but can also be seen in adults; and Miller-Fisher syndrome, which is a variant of Guillain-Barré syndrome that causes cerebellar ataxia and opthalmoplegia.

- **Acute Cerebellar Ataxia following head Trauma:**
Traumatic injuries to the cerebellum, brainstem or frontal lobe, producing interruption of the frontopontocerebellar tract can produce ataxia.

**Sensorial Ataxia:**

Disorders that damage the conscious proprioceptive pathway result in sensorial ataxia, which usually presents as long duration or gradually progressive ataxia. Among the main causes of sensorial ataxia are: multiple sclerosis (with affectation of posterior spinal columns or the medial lemniscus in the brainstem) (Fig. 16), vitamin B12 deficiency (which leads to subacute combined degeneration of spinal cord with demyelination of the posterior columns, posterior roots, and pyramidal tracts of spinal cord), vitamin E deficiency, Friedreich’s ataxia (Fig. 15), neurosyphilis (tabes dorsal which affects the lumbosacral dorsal columns and spinal nerve roots) and sensory polyneuropathies (diabetes mellitus, alcohol consumption).

**Vestibular ataxia:**

Vestibular ataxia can be caused by any focal lesion that affects the vestibular system including the vestibular areas of the cerebral cortex.

Vestibular ataxia can be caused by injuries to the vestibular nuclei in the brainstem or the VIII cranial nerve, either in the cerebellopontine angle or the internal auditory canal. Disorders affecting the vestibular nuclei include: tumors and ischemic or hemorrhagic strokes (Fig. 11 and 22); while cranial nerve damage can be produced by tumors (meningiomas and schwannomas) (Fig. 26), superficial siderosis (Fig. 27) or less frequently by meningeal carcinomatosis. Trauma with damage of vestibular structures can cause vestibular ataxia. Other causes of acute vestibular ataxia are labyrinthitis, vestibular neuronitis, labyrinthine infarction and perilymph fistula, all them of clinical diagnosis.

**Images for this section:**
Fig. 8: Metastases. 51 years old male with progressive ataxia. Left - Contrast enhanced computed tomography (CECT) demonstrates a cystic mass with ring enhancement. Magnetic resonance images (MRI) show a nodular solid-cystic mass in the right cerebellar hemisphere with hypointense signal on T1-weighted image (T1WI) (middle top), heterogeneous hyperintense signal with vasogenic peritumoral edema associated on T2- weighted (middle bottom) and FLAIR images (right top) and a ring pattern of enhancement on T1 with Gadolinium contrast (T1Gd)(right bottom). Metastases are the most frequent infratentorial tumors in adults.
**Fig. 9:** Medulloblastoma. 41 years old woman with headache, gait ataxia and dysarthria for two weeks. Left - Non-enhanced computed tomography (NECT) shows a nodular hyperdense mass in the right cerebellar hemisphere (due to its high nuclear-cytoplasmic ratio) The mass is hyperintense on T2 weighted image (T2WI) (middle top) with a solid pattern of enhancement on T1Gd (right top), and shows a restricted diffusion on diffusion weighted images (DWI) (middle and right bottom). This highly malignant neoplasm occurs more frequently at age 10 or before. Although much less common, it may also occur in adults, usually during the 3rd and 4th decades. In adults, medulloblastomas typically are located within the cerebellar hemispheres.
**Fig. 10:** Cerebellar Hemangioblastomas. Left. 47 years old woman with Von Hippel Hippel-Lindau syndrome with instability, dysarthria and dysmetria. There are postsurgical changes in posterior fossa with malacic cavities in right cerebellar hemisphere due to previous resection of hemangioblastomas and a typical cerebellar hemangioblastoma in the left cerebellar hemisphere: a cystic mass with a solid enhancing mural nodule (arrow). Right. 39 years old woman with gait ataxia. MRI shows a solid tumor in the right paravermian region, hypointense on T1WI (top), hyperintense on FLAIR and T2WI (middle) with a predominantly solid pattern of enhancement on T1Gd (bottom).
**Fig. 11:** 1. Low grade Astrocytoma of brainstem. 37 years old male with gait impairment and vertigo. MRI shows an ill-defined mass in the right dorsal pontomedullary junction, hyperintense on T2WI and FLAIR (top and middle) without enhancement after Gd administration (bottom). 2.- Low grade Astrocytoma of left thalamus. 35 years old male with gait ataxia. MRI shows a nodular mass in the left thalamus, hyperintense on T2WI and FLAIR (top and middle) and hypointense on T1WI (bottom). There was not enhancement on T1Gd (not shown).
Fig. 12: Extraaxial posterior fossa tumors as causes of cerebellar ataxia. 1. Meningioma. 53 years old woman with cerebellar ataxia. MRI shows an extra axial mass in the right pontocerebellar angle (PCA) isointense to gray matter on T2WI (top), hyperintense on FLAIR sequence (middle) with an avid enhancement on T1Gd (bottom). 2. Epidermoid cyst. 32 years old male with an incomplete previous resection of an epidermoid cyst in the left PCA and chronic gait impairment. MRI demonstrates a large mass in the left PCA with supratentorial extension and important mass effect against the middle cerebellar peduncle, the lateral pons and the cerebellar hemisphere on the left side. The mass shows a hypointense signal on T1WI (top), a hyperintense signal on T2WI (middle left), a heterogeneous isointense signal on FLAIR (middle right) and a restriction of diffusion on DWI (left and right bottom).
**Fig. 13:** Hindbrain malformations. Left. Dandy Walker Malformation. MRI shows a large cerebrospinal fluid (CSF) cyst posterior to the cerebellum which communicate with the 4th ventricle. The superior vermis is hypoplastic and rotated up over cyst while the inferior vermis is absent. Middle. Joubert Syndrome. CT demonstrates a 4th ventricle with "Bat-wing" shape (top), the midbrain shows a typically "Molar Tooth" shape (middle) and an abnormally deep interpeduncular fossa (bottom). Joubert syndrome is an inherited hypoplasia or aplasia of vermis. Clinically is characterized by oculomotor abnormalities, hypotonia, ataxia and usually variable mental retardation. Right. Rhombencephalosynapsis. MRI shows a fusion of cerebellar hemispheres and vermis agenesis, with typical transverse cerebellar folia and sulci (bottom). The patients of the left (19 years old male) and on the right (21 years old male) were diagnosed during their infancy and they both presented gait ataxia. The patient on the middle (33 years old female) showed mental retardation and gait ataxia, but there was not diagnosed until this CT that was done after an epileptic crisis.
**Fig. 14:** Spinocerebellar ataxia. 29 years old female with slowly progressive midline and symmetrical appendicular ataxia. MRI shows an important atrophy of cerebellar hemispheres and brainstem. There is a mega cisterna magna associated. There is also volume loss of cervical spinal cord.
**Fig. 15:** Friedreich’s ataxia. 19 years old female with proprioceptive impairment, gait instability and Romberg’s sign. Imaging findings include diminished cross-sectional area of the cervical spinal cord and medulla. There is a mild cerebellar volume loss.
Fig. 16: Demyelinating disorders - Multiple sclerosis. 36 years old woman with a two weeks sensorial ataxia. MRI demonstrates a lesion in the dorsal aspect of high cervical spinal cord. It is hypointense on T1WI (left) and hyperintense signal on T2WI (middle- sagittal plane and right top - axial plane). This lesion produces mass effect, and was diagnosed as a tumefactive demyelinating lesion. Axial FLAIR shows multiple hyperintense ovoid lesions in the periventricular white matter, suggestive of multiple sclerosis.
**Fig. 17:** Multiple system atrophy (MSA) cerebellar subtype. 77 years old female with three years of progressive ataxia associated to dysarthria an occasional urinary incontinence. There was not cognitive impairment. Sagittal T1WI (left) demonstrates marked pontine and cerebellar atrophy with an enlarged fourth ventricle. Coronal T1WI (top middle and right) shows a marked atrophy of cerebellar hemispheres and middle cerebellar peduncles. Axial T2WI (bottom middle and right) shows atrophy of pons and cruciform pontine hyperintensity known as "Hot cross bun" sign.
**Fig. 18:** Toxic Ataxia. 41 years old male with acute myelocytic leukemia and hematopoietic stem cell transplantation. The patient presented gait instability. Pharmacologic-toxic white matter lesion with hyperintense signal on T2WI (top) and FLAIR (left bottom) and mild increased diffusion on DWI (right bottom) in the periventricular and occipital white matter, without mass effect or "U" fibers affection.
Fig. 19: Central pontine myelinolysis. 61 years old alcoholic male with chronic gait ataxia. The clinical suspicion was Wernicke encephalopathy. MRI does not show any findings suggestive of Wernicke encephalopathy. Instead, it demonstrates a trident shape ("Trident" sign) central pontine hyperintense lesion on T2WI and FLAIR images (top) without mass effect. On T1WI (left bottom) the lesion shows low intensity signal with increased diffusion on DWI (right bottom). This findings suggest a chronic central pontine myelinolysis.
Fig. 20: Alcoholic Cerebellar Atrophy. 48 years old alcoholic male with chronic liver disease who presents with gait ataxia and dysarthria. MRI demonstrates a diffuse supratentorial and infratentorial atrophy, more accentuated in the posterior fossa with marked volume loss of vermis and cerebellar hemispheres.
Fig. 21: Postsurgical changes - Gliosis and Malacia. Malacic cavity with peripheral gliosis in the right cerebellar hemisphere with retraction of the fourth ventricle, secondary to previous resection of a meningioma. There is diffuse cerebellar atrophy associated.
**Fig. 22:** Acute Cerebellar Ataxia as a Possible Manifestation of Stroke. 1. Acute left lateral medullar infarct. 68 years old male with acute ataxia, vertigo, dysarthria, dysphagia and loss of pain and temperature sensation on the right side of the body and on the left side of the face (Wallenberg or lateral medullary syndrome). This is secondary to infarction in the left posterior inferior cerebellar artery (PICA) territory. MRI shows findings of an acute left lateral medullar infarct as a hyperintense focus on FLAIR (top) and T2WI (middle) on the dorsolateral region of the left medulla, with restricted diffusion on DWI (bottom). 2. Acute left paravermian cerebellar infarct. 82 years old male with gait ataxia. NECT shows a hypodense lesion in the left paravermian region.
Fig. 23: Acute Cerebellar Ataxia as a Possible Manifestation of Stroke. 1. 64 years old male with an accentuated headache, gait impairment, and instability. NECT shows a large hematoma on the right hemisphere and midline cerebellum with an important mass effect with total collapse of the fourth ventricle. It was an hypertensive hematoma. 2. 35 years old male with headache, gait ataxia and dysarthria. NECT demonstrates a hematoma on the left cerebellar hemisphere. Conventional cerebral angiography shows a pial arteriovenous malformation as the cause.
Fig. 24: Cerebellar abscess. 31 years old male with headache, fever and acute ataxia. MRI demonstrates two lesions located in the right middle cerebellar peduncle and in the left paravermian region (posterior to the fourth ventricle) with a high signal intensity on FLAIR (left top). The lesion in the left paravermian region shows a hyperintense center with a hypointense rim on T2WI (left top) and a ring pattern of enhancement on T1Gd (middle). Both lesions show a high signal (restricted diffusion) on DWI with a low ADC (left and right bottom, respectively). The final diagnosis was a meningitis with a secondary focus of cerebritis in the right middle cerebellar peduncle and an abscess on the left paravermian region.
Fig. 25: Spontaneous Creutzfeldt-Jakob disease. 86 years old female with gait ataxia and rapidly progressive dementia. MRI demonstrates a diffuse cerebral volume loss on sagittal T1WI (left top). Axial FLAIR image (left bottom) shows a bilateral symmetrical hyperintensity in heads of caudate nuclei, pulvinar nuclei ("Pulvinar" sign) and putamens. These hyperintense areas show restricted diffusion: low ADC (middle) and high signal intensity on DWI (right column).
**Fig. 26:** Right Vestibular Schwannoma. 73 years old male with right hearing loss and instability. Axial DRIVE sequence shows a mass in the right cerebellopontine angle with extension into the internal auditory canal (left) and avid enhancement on T1Gd images (right).
**Fig. 27:** Superficial Siderosis. Male patient with a saccular aneurysm in the anterior choroidal artery, ataxia and bilateral hearing loss. Axial FLAIR sequence shows a marked dark border on the surface of the brainstem (left) and cerebellopontine angles. Axial T2* GRE sequence (right) shows low signal along all brainstem, middle cerebellar peduncles and cerebellar surface. This occurs as a result of hemosiderin accumulation after recurrent subarachnoid hemorrhage.
Conclusion

The imaging evaluation of ataxia can be complex and confusing. Ataxia can be divided according to its clinical presentation in cerebellar, sensorial and vestibular ataxia. The most frequent form of presentation in all types of ataxia is the slowly progressive or long duration form, in which the most appropriated imaging procedure is head magnetic resonance. The acute presentation can be secondary to stroke, infection or head trauma. In suspected stroke or infection the preferred imaging procedure is again head magnetic resonance, if the treatment is not delayed. On the other hand, acute ataxia following head trauma must be evaluated with non-enhanced computed tomography to rule out hemorrhagic complications.

References