Clinical manifestations

Cerebellar ataxia

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Introduction
Definitions
The term ataxia (Greek, a (negative article); taxi (order) or lack of order) is often used synonymously with incoordination of movements. Ataxia is the most important sign of cerebellar disease, but there are other potential causes of ataxia. Cerebellar ataxia is defined as lack of accuracy or coordination of movement that is not due to paresis, alteration in tone, loss of postural sense, or the presence of involuntary movements [1]. Cerebellar ataxia relates to motor dysfunctions of the limbs, trunk, eyes, and bulbar musculature. Ataxia of gait refers to incoordination of walking. Postural ataxia refers to ataxia of stance and sitting, and includes truncal ataxia. Limb ataxia refers to incoordination of limb movements and ataxia of speech to cerebellar dysarthria. Postural and limb tremor are additional signs of cerebellar disease. Some, but not all authors think of tremor as a manifestation of ataxia.

Neurological findings in cerebellar disease
Patients with cerebellar disorders may walk with a wide-based, staggering gait, making it seem as if they were intoxicated by alcohol. A tendency to fall or deviate to one side suggests a unilateral cerebellar lesion on the same side. The stance is usually on a broad base with the feet several inches apart. In the mildest form, patients have difficulty standing with their feet together, in tandem position or on one foot. The Romberg sign might be present or absent in cerebellar disorders depending on the lesion site [2]. Several aspects of limb ataxia might be observed using the finger-to-nose and the heel-to-shin tests. Initiation of movement is delayed. The movement path of the limb is erratic and jerky (asynergia) and the nose is rarely touched at once (dysmetria). Subjects more often overshoot the target (hypermetria) and rarely stop the movement too soon (hypometria). Movement may be decomposed in time into its constituent parts (decomposition of movement). In patients with cerebellar strokes, rapid alternating movements are often irregular (dysdiadochokinesis) and slow (bradydiadochokinesis). The rebound test is frequently abnormal.

Neurological findings in cerebellar disease
Limb tremor occurs as a kinetic and, to a lesser extent, static tremor. Kinetic tremor occurs as an oscillatory movement when the patient initiates a movement of the limb or during the course of moving the limb. The tremor becomes more prominent as the moving limb approaches a target (intention tremor). There may be a rhythmic tremor of the body that can evolve into a severe titubation. Signs of cerebellar dysarthria include a slurred, monotonous and irregular speech with increased variability of pitch and loudness and articulatory impreciseness. The speech tempo is slow.

A variety of eye movement abnormalities are seen in cerebellar disease. Gaze-evoked nystagmus is a common finding. Downbeat nystagmus, upbeat nystagmus, and sustained horizontal nystagmus may also be present. Other frequent abnormalities are impairment of smooth pursuit and saccadic (ocular dysmetria) eye movements, inability to suppress the vestibular ocular reflex by fixation and abnormalities of optokinetic nystagmus.

Hypotonia, hyporeflexia, and asthenia were described as typical signs of acute, traumatic cerebellar lesions by Gordon Holmes [3].

Cognitive findings in cerebellar disease
Cerebellar involvement in a wide range of cognitive tasks as well as affect and behavior was proposed more than 20 years ago [4]. Human lesion studies suggest that the cerebellum may play a role in selected aspects of cognition, including frontal lobe function, language, and visuospatial processing. The right cerebellar hemisphere is thought to support language function, the left cerebellar hemisphere visuospatial function, and the vermis to modulate affect and behavior (“limbic cerebellum”) [5]. Findings appear to be more obvious in patients with acute cerebellar disorders, such as acute and subacute stroke. Because the posterolateral cerebellar hemispheres are considered supportive of cognitive functions, patients with lesions within the territory of the posterior inferior cerebellar artery (PICA) are at particular risk [6]. The role of the cerebellum in cognition, however, is still a matter of ongoing controversial discussion [7]. Frequently cited early findings, for example...
Disability of cerebellar patients in attention, have not been replicated in later studies or have been explained by motor components of the task. In addition to impaired motor function, it is unclear to what extent cognitive deficits are caused by nonspecific effects, such as accompanying hydrocephalus, depression, or global effects on brain metabolism following focal cerebellar lesions. Effects of extracerebellar lesions should also be considered. Although the cerebellum may be involved in certain cognitive tasks, the presenting symptoms of cerebellar disease are almost always motor.

**Associated neurological findings**
The presence of characteristic accompanying neurological symptoms and signs may help to localize the lesion within the territory of one of the cerebellar arteries or the cerebellar pathways.

Diplopia, facial numbness, facial droop, vertigo, or hearing loss resulting from associated cranial neuropathies (cranial nerves III, IV, V, VI, VII, VIII), Horner’s syndrome, and dysautonomia associated with ataxia suggest a disorder in the brainstem.

A slight loss of muscular power and increased fatigability of muscles may occur with acute cerebellar disorders. However, paresis associated with increased muscular tone, hyperreflexia, and extensor plantar reflexes (Babinski sign) suggest an additional involvement of upper motor neurons (corticospinal or pyramidal syndrome).

Pure cerebellar lesions do not cause disturbances in sensation. Hemianesthesia involving one side of the face, arm, and leg suggests an additional lesion of the sensory tracts (i.e., spinothalamic and medial lemniscal tracts) or contralateral parietal lobe. Frontal lobe disorders might cause cerebellar-like symptoms with walking difficulties and clumsiness. Frontal lobe lesions are commonly associated with impairment of cognitive function and changes in personality, and often cause urinary incontinence.

**Anatomy of the cerebellar system**

**Cerebellar subdivisions**

There are two gross anatomical subdivisions of the cerebellum [8]. First, the cerebellum can be subdivided into two lateral parts, called the hemispheres, and a midline structure, the vermis (Figure 3.1(a), right side). Usually, the two flocculi are considered separately. Second, based on the main fissures, the cerebellum is subdivided into three lobes: the anterior, the posterior, and the flocculonodular lobe (Figure 3.1(a), left side). The anterior and posterior lobes are further subdivided into smaller lobules. The terminology used to identify the lobules is not uniform. Several nomenclature systems use individual names. For example, the cerebellar tonsil is the most caudal lobule of the hemispherical part of the posterior lobe. Larsell introduced a numbering system, which consists of Roman numerals in the vermis (I-X) and the prefix H in the hemispheres. Today a modified version of the Larsell terminology is frequently used, proposed by Schmahmann and coworkers [9].

The terms archicerebellum, paleocerebellum, and neocerebellum originate from phylogenetic and embryological studies. The terms vestibulo-, spino-, and cerebrocerebellum originate from termination sides of cerebellar afferent projections. These subdivisions match well with the subdivisions based on phylogenetic studies. The flocculonodular lobe (~ archicerebellum) has been named vestibulocerebellum because of heavily projecting vestibular afferents; the vermis and paravermal parts of the cerebellar hemispheres (~ paleocerebellum) were called spinocerebellum because of their spinal afferents, and the cerebellar hemispheres (~ neocerebellum) cerebrocerebellum (synonym pontocerebellum) based on their corticopontine input (Figure 3.1(a), right side).

On the basis of the efferent projections from the cerebellar cortex to the cerebellar nuclei, Jansen and Brodal [10] suggested a subdivision into three longitudinal (sagittal) zones: a medial zone (vermis) projecting to the fastigial nucleus, an intermediate zone (paravermal part of the cerebellar hemisphere) projecting to the interposed nuclei (i.e., globose and...
The main connections of the dentate and interposed nuclei. Note that the ascending connections of the cerebral cortex from the dentate nucleus are synaptically interrupted in the thalamus. Both the spinal cord (via the red nucleus) and the cerebral cortex (via the thalamus) can be influenced via the interposed nucleus (emboliform and globose nuclei in humans). (With permission from Brodal P. *The Central Nervous System. 2nd edn. New York, NY: Oxford University Press, 1998; 400, 402.)

**Figure 3.2.** The main connections of the dentate and interposed nuclei. Note that the ascending connections of the cerebral cortex from the dentate nucleus are synaptically interrupted in the thalamus. Both the spinal cord (via the red nucleus) and the cerebral cortex (via the thalamus) can be influenced via the interposed nucleus (emboliform and globose nuclei in humans).

Emboliiform nuclei, and a lateral zone (lateral part of the cerebellar hemisphere) projecting to the dentate nucleus (Figure 3.1(a), right side). Later studies showed that the longitudinal subdivision was more detailed.

**Cerebellar pathways**

The cerebellum is connected with the brainstem by afferent and efferent fibers passing through three pairs of tracts, called the inferior, middle, and superior cerebellar peduncle (or restiform body, brachium pontis, and brachium conjunctivum). The middle cerebellar peduncle contains only afferent fibers. In the inferior peduncle most fibers are afferent, whereas in the superior cerebellar peduncle most fibers are efferent.

The cerebellar cortex receives afferent input from most parts of the peripheral (proprioceptive, cutaneous, vestibular, visual, and possibly olfactory) and central nervous system [11]. From the trunk and legs, the dorsal and ventral spinocerebellar tracts enter the cerebellum through the ipsilateral inferior and contralateral superior cerebellar peduncle, respectively. From the arms and neck, the cuneo- and rostral spinocerebellar tracts enter the cerebellum through the inferior and superior cerebellar peduncles. Many afferent pathways have additional relay stations (pontine nuclei, inferior olives) before they enter the cerebellum. From the inferior olive, climbing fibers enter the cerebellum through the contralateral inferior cerebellar peduncle. The pontine nuclei represent the most important relay for corticocerebellar pathways. Corticopontine projections enter the cerebellum mainly through the contralateral middle cerebellar peduncle.

The cerebellar nuclei are the principal source of cerebellar efferent fibers. Efferent cerebellar pathways descend to the brainstem and spinal cord and ascend to the cerebral cortex. Efferents from the flocculonodular lobe project mainly to the vestibular nuclei in the brainstem directly and indirectly via the fastigial nuclei. Efferents from the globose and emboliiform nuclei form the major cerebellar projection to the contralateral red nucleus (cerebellorubral tract). The main efferent output from the red nucleus projects to the spinal cord on the contralateral side (rubrospinal tract). Most fibers from the dentate nuclei end in the contralateral thalamus. The main projection from the thalamus goes to motor and premotor cortices via the internal capsule (Figure 3.2) [12]. Efferent projections, however, have also been shown to prefrontal and parietal areas [6]. Because the ascending fibers from the cerebellum to the red nucleus and the motor cortex and the descending fibers from the red nucleus and cerebral cortex to the spinal cord are crossed, the cerebellar hemisphere exerts its influence on the body half of the same side. Therefore, in unilateral cerebellar lesions, symptoms of limb ataxia occur ipsilaterally.

**Afferent connections from the cerebral cortex**

The vast majority of the afferents to the pontine nuclei arise in the cerebral cortex and form the corticopontine tract. The corticopontine tract is uncrossed whereas most of the pontocerebellar fibers cross; thus the cerebral cortex of one side acts mainly on the cerebellar hemisphere of the opposite side. A large portion of the corticopontine fibers arise in the primary motor (M1) and sensory (S1) cortices. There are substantial contributions also from the supplementary motor area (SMA) and premotor area (PMA) as well as from areas 5 and 7 of the posterior parietal cortex. The pontine nuclei also receive afferents from prefrontal areas 8, 9, and 46, the visual cortex, and parts of the hypothalamus and limbic structures [6, 13].

The corticopontine tract runs in the internal capsule and in the crus cerebri. The frontopontine tract (Arnold’s bundle) is localized in the anterior limb of the internal capsule, the parietopontine tract in the posterior limb, the occipitopontine tract in the retrolenticular portion of the posterior limb, and the temporopontine tract ( Türk’s bundle) in the sublenticular portion of the posterior limb of the internal capsule. The frontopontine tract lies near the corticobulbar tract located in the genu of the internal capsule and the corticospinal tract located in the anterior part of the posterior limb (Figure 3.3(a)). The internal capsule turns rostrally into the corona radiata and caudally into the crus cerebri. In the crus cerebri, the frontopontine tract is localized medially and the temporopontine tract...
lateral to the corticospinal tract (Figure 3.3(b)). The smaller parieto- and occipitopontine tracts are found medial to the temporopontine tract. The corticopontine tract remains in close relationship to the corticospinal and corticobulbar tracts on their way through the mesencephalon and the base of the pons (Figure 3.4).

**Efferent connections to the cerebral cortex**

The fibers from the dentate nucleus leave the cerebellum through the superior cerebellar peduncle. They cross the midline in the mesencephalon, and some fibers end in the contralateral red nucleus. Most fibers continue rostrally, however, to end in the thalamus, primarily in the ventrolateral (VL) nucleus. Some also reach the ventroanterior (VA) nucleus. Thalamic fibers from the VL and VA nuclei course through the superior thalamic peduncle in the posterior limb of the internal capsule to the precentral region (M1, SMA, and PMA). In the internal capsule, the corticospinal tract lies in close vicinity to the thalamic fibers (Figures 3.2 and 3.3).

The sensory tracts do not travel close to the cerebellar fibers until they reach the thalamus. The spinothalamic tract and the medial lemniscus, however, do travel in close proximity to the superior cerebellar peduncle in the superior lateral pons, and when affected, result in a crossed hemisensory ataxic syndrome. The sensory tracts synapse in the
ventroposterolateral nucleus (VLP) of the thalamus and run through the superior thalamic peduncle to the postcentral region. Therefore, sensory and cerebellocortical fibers are in close proximity in the thalamus and the posterior limb of the internal capsule (Figure 3.5).

Dum and Strick distinguish between a so-called motor and a nonmotor domain of the dentate nucleus [14]. According to their anatomical findings, projections to the PMA and to M1 in the dorsal portion of the dentate map resemble the motor domain of the dentate nucleus. The nonmotor domain in the ventral portion of the nucleus contains output channels that project to prefrontal and parietal areas concerned with executive (areas 46d, 9l) and visuospatial functions (area 7b). Connections to nonmotor cortical areas are modest compared to those reaching motor areas.

Vascular supply of the cerebellum

The vascular supply of the cerebellum is provided by three arteries, the superior cerebellar artery (SCA), the posterior inferior cerebellar artery (PICA), and the anterior inferior cerebellar artery (AICA) [15]. The cerebellar vascular territories are well illustrated in Tatu et al. [16], based on pathological studies by Amarenco [17] and the injection studies by Marinkovic et al. [18]. Variations in the size and distribution of these vessels are frequent, and all major branches are highly anastomotic. All cerebellar arteries supply cerebellar as well as brainstem structures. Therefore, vascular disorders may damage the cerebellum and brainstem together.

The SCA supplies the superior parts of the cerebellum down to the horizontal fissure (lobules I to Crus I, which is part of lobule VII). It supplies all cerebellar nuclei, most of the cerebellar white matter, and the superior cerebellar peduncle. The SCA supplies the dorsolaterotegmental area of the upper pons. Branches of the PICA supply the inferior aspect of the cerebellar hemispheres and inferior vermis extending up...
to the horizontal fissure (lobules X to Crus II, which is part of lobule VII). The PICA occasionally supplies posterior inferior parts of the dentate nucleus and it probably supplies the fastigial nucleus as well. The medial branch of PICA supplies a part of the dorsal medullary area.

The SCA and PICA have two main branches, supplying the more dorsomedial (m) and the more anterolateral (l) parts of each territory. Medial branches supply mostly the vermis and paravermian parts of the hemispheres, and lateral branches the more lateral parts of the hemispheres. Branches of the AICA supply the flocculus, adjacent lobules of the inferior and anterior cerebellum, and the middle cerebellar peduncles. The AICA supplies the lower third of the lateral pontine territory in most individuals, its middle third frequently, and the superior part of the lateral region of the medulla oblongata in a few cases. The labyrintheine artery, which supplies the inner ear, arises frequently from the AICA. (For further details and diagrammatic drawings, see Section 2, Chapter 41: Cerebellar infarcts.)

**Cerebellar syndromes**

Gross anatomical subdivision in the hemispheres and vermis gives a first idea of functional localization within the cerebellum. The vermis is involved in the control of balance and eye movements, and the hemispheres are involved in limb coordination. Lesions of the vermis result in disturbances of stance, gait, and ocular movements, whereas lesions of the cerebellar hemispheres mostly result in ipsilateral limb ataxia and dysarthria.

Three clinical syndromes of cerebellar dysfunction are commonly distinguished [19]. Lesions of the flocculonodular lobe cause postural ataxia of the trunk during sitting, standing, and walking (flocculonodular syndrome). Patients frequently fall even during sitting. In patients with such lesions, severe postural sway is present with eyes open and is essentially unchanged with eyes closed (absence of Romberg’s sign). Fine coordinated movements of the limbs are commonly unimpaired. Saccadic slow pursuit, nystagmus, and an inability to suppress the vestibuloocular reflex (VOR) are often present.

Damage to the vermal part of the anterior lobe results in ataxia of stance and gait (anterolobe syndrome). Patients with this disorder develop a severe disturbance of standing and walking, with relatively preserved fine coordinated movements of the upper limbs. Lesions lead to anteroposterior body sway with a frequency of about 3 Hz. Visual stabilization of posture is preserved and the tremor is provoked by eye closure (presence of Romberg’s sign).

Lesions of the cerebellar hemisphere are followed by ipsilateral limb ataxia including hypotonia in acute lesions, and if the dentate nucleus is involved, kinetic tremor (neocerebellar syndrome). Past pointing and deviation of gait to the affected side are associated symptoms. Limb ataxia is primarily a consequence of a lesion of the intermediate zone (paravermian cortex projecting to the interposed nucleus). Two inverted somatotopic maps have been charted for the cerebellar cortex [20]. The leg is represented anteriorly within the anterior lobe, with the arm and face represented successively more posteriorly. In the posterior lobe, the arrangement is the reverse, with the face represented anteriorly.

Lesions of the superior paravermian cerebellum are accompanied by dysarthria. Early findings that left hemispherical cerebellar lesions appear to be more frequently associated with cerebellar dysarthria than right hemispherical lesions have not been replicated by later studies [21].

Schmahmann and Sherman [5] have proposed the “cerebellar cognitive affective syndrome” as a newly defined clinical entity. According to their findings, patients with lesions involving the posterior lobe of the cerebellum including the vermis, but not the anterior lobe, present with behavioral abnormalities. These changes were characterized by: impairment of executive functions such as planning, set-shifting, verbal fluency, abstract reasoning, and working memory; difficulties with spatial cognition including visuospatial organization and memory; personality change with blunting of affect or disinhibited and inappropriate behavior; and language deficits including agrammatism and dysprosodia. At present, however, it is a matter of ongoing discussion whether cognitive dysfunction contributes to the clinical picture of cerebellar disease.

In spite of the prototypic descriptions, cerebellar syndromes in practice often reflect more than one of the functionally different subdivisions of the cerebellum. Furthermore, there is no difference between the symptoms of ataxia that result from lesions of afferent and efferent cerebellar pathways and those that result from the cerebellum itself. A remarkable degree of compensation occurs in lesions of the cerebellum, which is most prominent in lesions restricted to the cerebellar cortex. A comparison of patients with acute and chronic focal cerebellar lesions due to stroke or tumor surgery showed that lesions of the cerebellar nuclei, but not the cerebellar cortex, were followed by remaining motor signs regardless of the age at injury and the pathology [22].

**Vascular syndromes resulting in ataxia**

Vascular lesions of the cerebellum itself and of the corticopontocerebellar and dentatothalamic pathways might result in ataxia.

**Cerebellar infarcts**

First with the introduction and later with better availability and quality of CT and MRI scanners, the number of detected cerebellar infarcts has increased. It became clear that the classical cerebellar ischemic syndromes, including brainstem signs as well as life-threatening brainstem compression and hydrocephalus from postinfarct edema, are comparatively rare [23,24]. The majority of cerebellar infarctions have a benign clinical course [25].

Infarcts of the SCA and PICA are most common; AICA infarcts are rare [26]. Whereas PICA and SCA infarcts are
frequently restricted to the cerebellum, AICA territory infarcts almost always include the brachium pontis and the lateropontine area, and brainstem signs predominate [27–29]. Symptoms and signs differ depending on the vascular territory and are in good accordance with the known functional compartmentalization within the cerebellum [19]. Infarction in the territory of any of the three cerebellar arteries results in limb and gait ataxia [23,30]. Dysarthria is a characteristic finding in SCA distribution infarcts, whereas vertigo is particularly common in infarcts in the PICA and AICA territories [23,31,32].

During the first stage after a cerebellar infarct, goal-directed movements are hypermetric due to a delayed onset of the antagonist activity. Both the intensities of the agonist and antagonist are depressed. As a result, patients initially complain of weakness [33]. Abnormal reprogramming of the electromyographic (EMG) pattern may result in the shifting from hypermetria to hypometria [34].

**Posterior inferior cerebellar artery**

In instances of combined cerebellar and medullary infarction in the PICA territory, the patient has the features of Wallenberg’s lateral medullary syndrome: ipsilateral limb ataxia and gait imbalance is accompanied by vertigo, nystagmus, occipitocervical headache and ipsilateral facial pain, dysphonia and dysphagia, ipsilateral facial loss of pain and temperature sensation, Horner’s syndrome, palatal weakness, and contralateral thermalalgesia of the limbs and trunk. Ipsilateral lateropulsion may be present. All these features can be present in full and can occur in various combinations. Brainstem involvement, however, is rare [23,24,35].

Only about one in five PICA cerebellar infarcts are accompanied by infarction in the lateral medulla oblongata. Cerebellar infarcts in the PICA territory that spare the dorsolateral medulla oblongata present with occipitocervical headache ipsilateral to the infarct, along with acute vertigo, nausea and vomiting, nystagmus, ipsilateral limb ataxia, and gait ataxia [23,24,29].

Oculomotor signs (nystagmus) and ataxia of stance and gait are more common in infarction of the medial compared to the lateral branches of the PICA [17,35,36]. Limb ataxia is a sign of infarction in the territory of the lateral branches of the PICA. Limb ataxia in PICA infarction has been related to additional damage to branches of the proximal intracranial territory supplying the inferior cerebellar peduncle, one of the important input structures of the cerebellum, or to damage in the relevant output structure, the interposed or dentate nuclei [37]. This would imply that lesions of the body representation in the posterior lobe of the cerebellum are not followed by limb ataxia.

Initial hoarseness has been described, but not dysarthria [38]. Isolated vertigo and gait imbalance without other manifestations may be present, which resembles the presentation of labyrinthitis [39]. This syndrome is most common in infarcts in the territory of the medial branch of the PICA. In labyrinthitis, however, the direction of nystagmus is independent of the direction of gaze, whereas in PICA infarction gaze-evoked nystagmus is primarily direction changing.

**Superior cerebellar artery**

A unilateral, isolated, full SCA infarct, including the pontine territory, is rare. It results in ipsilateral limb ataxia, Horner’s syndrome, choreiform involuntary movements, contralateral thermalalgesia, and fourth cranial nerve palsy [27]. Most SCA territory infarcts affect only fractions of the territory. A common presentation is the acute onset of gait imbalance and ipsilateral limb ataxia sometimes associated with headache, vertigo, nausea, and vomiting [40,41]. Axial lateropulsion occurs. Vertigo, however, is less common than in PICA- and AICA-distribution infarcts. The relatively lower frequency of vertigo in SCA territory infarct patients has been related to the comparatively fewer vestibular connections of those portions of the cerebellum supplied by the SCA, in contrast to the rich connections of the flocculonodular lobe supplied by the PICA and AICA [23]. Horizontal nystagmus is present in at least 50% of patients. Dysarthria is a prominent feature [32,38]. Limb ataxia appears to be more prominent in SCA cases as compared to PICA distribution infarcts [24,30].

Dysarthria and ataxia of stance and gait are more severe in infarction of the medial compared to the lateral branches of the SCA [41,42]. Limb ataxia is a sign of infarction of the lateral branches of the SCA. In addition, SCA infarcts can be accompanied by other infarcts in the territory of branches of the rostral basilar artery. Symptoms of rostral brainstem and occipital involvement may overshadow signs of cerebellar involvement [27,43].

**Anterior inferior cerebellar artery**

In PICA and SCA territory infarcts, the clinical signs are dominated by cerebellar infarction, while in AICA territory pontocerebellar infarcts, brainstem signs predominate [28]. An almost complete syndrome is observed in most cases. The classical syndrome of AICA occlusion involves vertigo, tinnitus, ipsilateral hearing loss, dysarthria, peripheral facial palsy, Horner’s syndrome, multimodal facial hypesthesia, and ipsilateral limb ataxia accompanied by contralateral thermalalgesia of the limbs and trunk. Partial AICA syndrome can be confused with Wallenberg’s syndrome. The prominence of auditory involvement and peripheral facial palsy point to a clinical diagnosis of AICA territory infarction. Partial AICA syndromes rarely can present with pure vertigo (mimicking labyrinthitis) or isolated ipsilateral ataxia [28,44].

**Lacunar stroke of the pons and internal capsule: ataxic hemiparesis**

Vascular lesions in the course of the corticopontocerebellar and dentatothalamic pathways may result in ataxia, primarily of the limbs. Because of the close vicinity of the cerebellar...
pathways and the corticospinal tracts in the base of the pons, crus cerebri, and internal capsule, hemiataxia is often accompanied by homolateral pyramidal signs (Figures 3.3 and 3.4).

The vascular syndrome of ataxic hemiparesis (i.e., hemiparesis and ataxia involving the limbs on the same side) was first named by Fisher in 1978 [45]. Incoordination is out of proportion to weakness. Ataxic hemiparesis has been found to be highly predictive of a lacunar lesion [46]. The syndrome, however, does not predict the lesion location. Rather, in ataxic hemiparesis infarcts are scattered throughout the motor pathway. The most common lesion sites are the posterior limb of the internal capsule (23%–44%) and pons (19%–31%). Other lesion sites are the thalamus, anterior part of the corona radiata, basal ganglia, and frontal subcortical white matter [46,47]. Although the most common lesion in all studies was a small deep infarct, cerebral hemorrhage and superficial infarcts in the cerebellum (SCA territory) and frontal cortex (anterior cerebral artery territory) have also been shown to cause ataxic hemiparesis in a small percentage of cases [46–48].

The clinical features of ataxic hemiparesis with different locations are almost identical. Ataxia nearly always involves the arm and leg with equal intensity. Minor associated signs are common, such as paresthesiae with thalamic infarction, and dysarthria, nystagmus, and gait ataxia with a pontine infarct [47–49]. Most investigators believe that the syndrome of ataxic hemiparesis is due to simultaneous involvement of corticospinal and dentatorubrothalamicocorticopontocerebellar pathways. The clinical significance of accompanying cerebellar diaschisis remains unknown. Cerebellar hypoperfusion and hypometabolism is a common but not mandatory finding following lesions within the cerebral cortex and thalamus (crossed cerebellar diaschisis; [50,51]) and the brainstem (uncrossed and crossed cerebellar diaschisis; [52,53]). Newer studies suggest a possible relationship between lacunar stroke, diaschisis, and recovery potential [54].

**Internal capsule**

The posterior limb of the contralateral internal capsule is a common lesion site in ataxic hemiparesis, sometimes extending toward the corona radiata region and lateral thalamus [46,47,55]. In these cases, the frontopontine and temporoparietopontine bundles are not involved, because they course in the anterior limb and retro- or sublenticular portion of the internal capsule, respectively (Figure 3.3). The cerebellar dysfunction is considered to be related to destruction of corticopontine fibers from the precentral cortex or connecting fibers between the ventrolateral nucleus of the thalamus and the precentral region (i.e., superior thalamic tract) [56]. Saitoh et al. [57] studied ataxia and the readiness potential in four cases of ataxic hemiparesis resulting from a small infarct in the posterior limb of the internal capsule. On the basis of normal readiness potentials, the dentatothalamocortical system, secondary to interruption of the thalamic radiation at the internal capsule, did not appear to be significantly involved. Saitoh et al. concluded that the ataxia appeared to be the result of involvement of the corticopontine tract originating from the precentral region (areas 4 and 6) at this level.

Ataxic hemiparesis is frequently accompanied by hemisensory signs because the sensory tracts course within the superior thalamic tract in the posterior limb of the internal capsule (Figure 3.3) [55].

**Pons**

Bassetti et al. [58] delineated three main syndromes of isolated infarcts of the pons (ventral, tegmental, and bilateral). Ventral lesions interrupt the corticospinal, corticobulbar, and corticopontine fibers as well as pontine neurons to various extents (Figure 3.4). Small focal lesions of the ventral pons result in distinct syndromes: ataxic hemiparesis, dysarthria–clumsy hand syndrome, dysarthria–dysmetria and dysarthria–facial paresis [59,60]. These syndromes, which are not absolutely discrete, reflect the topography of motor organization in the basis pontis (i.e., anterior or ventral pons) [59].

Eye movement disorders, sensory disturbances, and cranial nerve (V–VII) dysfunction suggest additional tegmental pontine infarction. Whereas ventral pontine lesions result in limb ataxia, medial tegmental lesions are followed by ataxic gait [61]. Occasionally, bilateral infarcts can be limited to the ventral pons, presenting with ataxic tetraparesis, almost isolated para- and tetraplegia, or locked-in syndrome [58].

**Thalamus**

Hemiataxia is a common occurrence in thalamic infarction involving the ventrolateral part of the thalamus, usually from involvement of the thalamogeniculate territory [62–64]. Hemiataxia as a manifestation of thalamic infarction rarely occurs in isolation, being associated with ipsilateral hemiparesis (ataxic hemiparesis), pain and hemiparesis (painful ataxic hemiparesis), ipsilateral sensory disturbance (hemiataxia–hypesthesia), and ipsilateral sensory disturbance and hemiparesis (hypesthetic ataxic hemiparesis). These four syndromes might be explained by variations in the blood supply to the capsulothalamic region [47]. Accompanying sensory signs are frequent in thalamic hemiataxia because of the close neighborhood of the sensory (i.e., VPL) and cerebellar (i.e., VL and VA) thalamic nuclei in the ventral thalamic group (Figure 3.5). The occurrence of pain has localizing value. Pain is not present in pontine, mesencephalic, or capsular ataxic hemiparesis [62].

In contrast to the ascending dentatothalamic and sensory pathways, the corticospinal tract is not part of the thalamus. Corticospinal signs in thalamic ataxic hemiparesis are probably due to ischemia or edema compressing the adjacent corticospinal tract or associated infarction of the adjacent internal capsule, as the thalamogeniculate branches sometimes contribute to the innermost part of the posterior limb of the internal
Subcortical white matter lesions

Abnormalities of gait are common in subcortical vascular encephalopathy [66,67]. The gait pattern in subcortical vascular encephalopathy shows similarities to that of some patients with hydrocephalus, frontal lobe lesions, and senile disorders of gait [46]. The gait disorder has elements of both parkinsonism and cerebellar ataxia. Patients stand on a wide base, are slightly ataxic, and take slow and shuffling parkinsonian steps [66]. Patients show relatively preserved function of the upper limbs, lively facial expression, and less flexed posture, which has led to the use of the term lower body parkinsonism (marche à petits pas, arteriosclerotic parkinsonism) [69]. Frequently associated signs are cognitive impairment and minor gait initiation failure may remain the sole symptom for many years (isolated gait initiation failure or Petren’s gait). On the other hand, the clinical picture may be dominated by impairment of balance (e.g., truncal ataxia), severe enough to prevent standing and walking (frontal dis-equilibrium) [67].

A single-photon emission computed tomography (SPECT) study in patients with subcortical vascular encephalopathy showed hypoperfusion of the SMA and cerebellum [70]. Ischemia/disconnection between the basal ganglia and SMA, and between premotor areas and the cerebellum may explain alterations in gait [66,71].

Cortical strokes

Cortical strokes can be followed by ataxic hemiparesis. Large infarcts of the anterior cerebral artery have been found to cause leg-predominant weakness with ipsilateral ataxia (homolateral ataxia and crural paresis) [46,56]. In addition, small cortical lesions of the precentral gyrus have been shown to result in ataxic hemiparesis [46]. These findings are explained by the afferent and efferent connections of the primary motor cortex to the cerebellum.

Concluding remarks

Limb ataxia and ataxia of gait are common in SCA, PICA, and AICA territory infarctions. Dysarthria is a characteristic symptom of stroke in the SCA distribution. Vertigo as a presenting sign is most common in PICA and AICA territory infarcts. Concomitant brainstem infarction is the rule in patients with infarcts in the AICA distribution, but not in the SCA or PICA territories.

Ataxic hemiparesis occurs most commonly in lacunar lesions of the internal capsule and base of the pons. Accompanying sensory signs and pain suggest a lesion site in the thalamus and dysarthria suggests a lesion in the pons. Homolateral ataxia with crural paresis is the result of superficial anterior cerebral artery infarction. Gait disorders in subcortical arteriosclerotic encephalopathy show parkinsonian and ataxic features.

References

Section 1: Clinical manifestations

Berlin, Germany: Springer-Verlag, 1985; 126–147.


56. Bogousslavsky J, Martin R, Moulin T. Homolateral ataxia and crural paresis:


