Ataxia

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Abstract
Ataxia is derived from a Greek word meaning 'lack of order' and can be defined as impairment of coordination in the absence of significant muscle weakness. Patients complain of poor balance when walking and sitting, difficulties with accurate hand movements, tremor and slurred speech. In many cases, the presence of ataxia indicates pathology in the cerebellum and/or its brainstem connections, or an impairment of sensory information. Although the differential diagnosis of ataxia is complex, a careful history and examination can reduce the number of possible causes. This article discusses the principal features, investigation and management of ataxia caused by cerebellar disease and, to a lesser extent, sensory neuropathies in which ataxia is a prominent feature.

Keywords ataxia; cerebellum; Creutzfeldt—Jakob disease; Friedreich's; multiple sclerosis; paraneoplastic; proprioception; spinocerebellar ataxia

Types of ataxia

Cerebellar ataxia

Cerebellar ataxia indicates pathology in the cerebellum and/or its brainstem connections. The cerebellum is located in the posterior fossa and is linked to the brainstem and cerebrum via three pairs of cerebellar peduncles (inferior, middle and superior). It comprises two hemispheres and a central region termed the 'vermis'. How the cerebellum co-ordinates complex fine movement is unclear, but it is thought to balance the activity of agonist and antagonist muscles to smooth out voluntary movements, acting on information from various sources, including the vestibular system, joint proprioception and muscle tone. The cardinal features of cerebellar disease are illustrated in Box 1. Disease affecting one cerebellar hemisphere causes ataxia in the ipsilateral limbs, whereas damage to the vermis predominantly affects gait and sitting balance, with relative sparing of eye movements and speech.

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What's new?

- The study of genetic ataxias has progressed rapidly. The phenotype of existing syndromes has widened and new protein mutations have been implicated
- Although most cerebellar ataxias are untreatable, some (e.g. inflammatory causes, vitamin E deficiency) may respond to treatment and therefore need to be considered
- Variant Creutzfeldt—Jakob disease is less prevalent now since new public heath measures have been implemented
- Specific brain MRI abnormalities are recognized with many causes of cerebellar ataxia

Sensory ataxia

This results from pathology affecting the proprioceptive system anywhere from the peripheral sensory nerves, via the dorsal root ganglia and the dorsal columns in the spinal cord to the medulla. It can be distinguished from ataxia caused by cerebellar disease by demonstrating impaired joint position sense and unsteadiness that is significantly worse with the eyes closed (positive Romberg's test). Sensory ataxia is not typically associated with dysarthria or nystagmus.

Vestibular ataxia

Vestibular disease is often associated with vertigo, nystagmus and gait ataxia, but testing of individual limbs reveals no limb ataxia.

Causes of ataxia

The three types of ataxia have overlapping causes, and therefore can either co-exist or occur in isolation. This review will concentrate mainly on the causes of cerebellar ataxia.

Cardinal features of cerebellar disease

- Clumsiness of the hands when performing rapidly alternating movements such as pronation and supination of the forearm (dysdiadochokinesis)
- Jerky over-shooting of a limb as it approaches a target (e.g. finger—nose testing) (intention tremor)
- Unsteady, wide-based gait (in its most subtle form, may manifest as difficulty walking heel-to-toe)
- Impaired coordination on heel—shin testing
- Slurred, dysarthric speech (may have a 'scanning' quality, in which speech is broken into syllables)
- Abnormal eye movements (e.g. nystagmus, jerky pursuit eye movements)
- Muscle hypotonia

Box 1

Ataxia of acute onset (onset minutes/hours)

Vascular disorders: sudden-onset hemi-ataxia, particularly if associated with vertigo, vomiting and brainstem signs, is strongly suggestive of an infarct or haemorrhage affecting the cerebellum or its brainstem connections (Figure 1). The history may reveal risk factors such as hypertension and vascular disease. This is a neurological emergency as it may result in hydrocephalus by occluding cerebrospinal fluid (CSF) flow. Urgent CT scanning and neurosurgical referral are indicated in such cases.

Intoxication: alcohol is one of the most common causes of acute and chronic cerebellar dysfunction and vestibular ataxia. Ataxia due to intoxication is usually bilateral and often involves the trunk/gait. Other cerebellar symptoms and signs are less common. Altered mentation is often also present and, if combined with complex eye movement abnormalities, suggests Wernicke's encephalopathy (due to thiamine deficiency). The diagnosis can be confirmed by measuring red blood cell (RBC) transketolase, but this should not delay treatment with parenteral thiamine.

Drugs, such as anti-epileptic medications, lithium and some recreational drugs (e.g. ketamine, phencyclidine (PCP)), cause ataxia in a predictable, dose-dependent manner. Drug toxicity, confirmed by measuring drug concentrations in plasma/serum, should always be considered in a patient with epilepsy or manic depression who presents with cerebellar signs.

Migraine: unsteadiness/dizziness can occur in typical migraines. In addition, the rare basilar migraine variant can present with transient cerebellar ataxia and brainstem signs; headache may not be prominent.

Ataxia of subacute onset (hours/days)

Infections: viral cerebellitis and rhomboencephalitis are the most common causes of subacute ataxia in children, especially those aged 2–10 years. Usually recovery occurs over a period of weeks. Post-infectious encephalomyelitis may also occur, especially related to varicella infection.

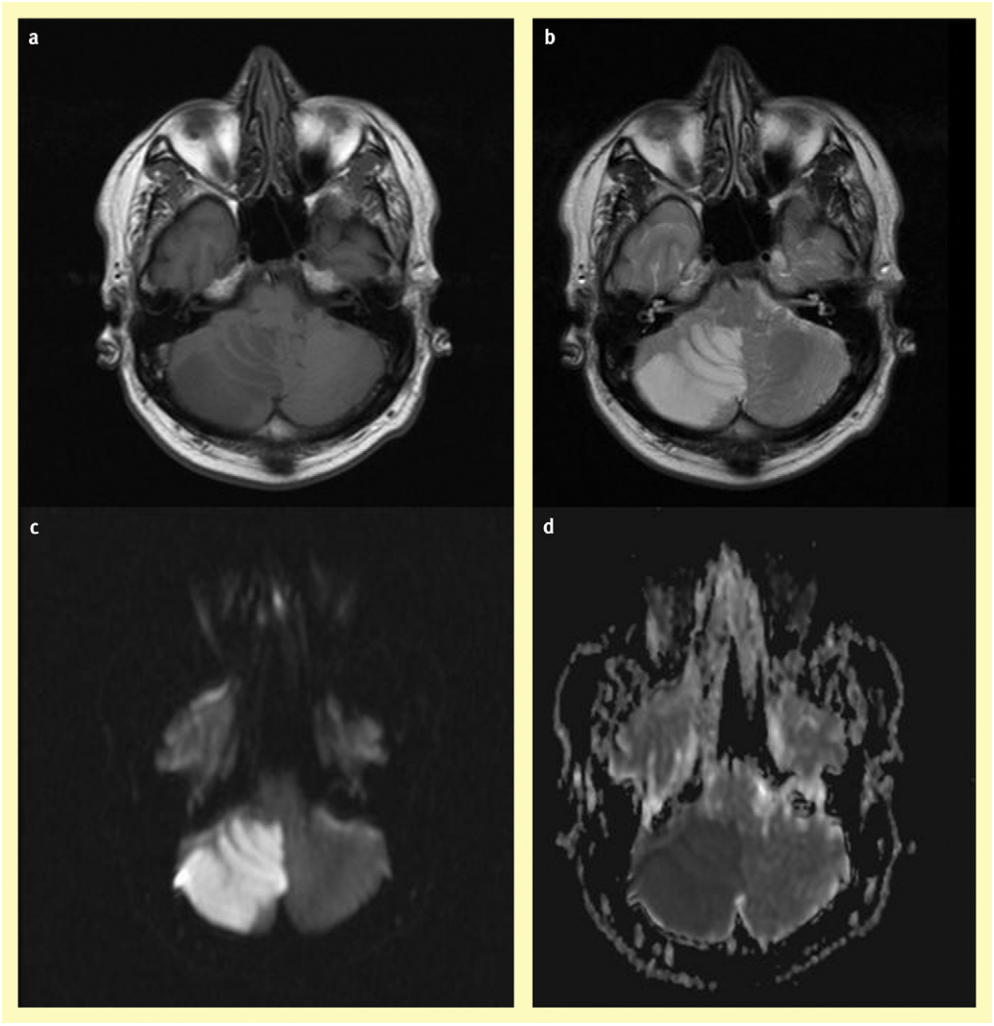

Figure 1 MRI brain scan of a 36-year-old man who presented with sudden-onset headache, vomiting and ataxia. The axial images reveal an acute right posterior inferior cerebellar artery (PICA) territory infarct. (**a**) T1-weighted image, (**b**) proton density (PD) + T2-weighted image, (**c** and **d**) diffusion-weighted images illustrating restricted diffusion in the right cerebellar hemisphere.