Ataxia of parietal lobe origin

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Ataxia is the cardinal sign of cerebellar dysfunction and when it occurs, it indicates the possibility of cerebellar lesions. However, this may not always be the correct explanation. Appenzeller and Hanson[1] reviewed studies, published between 1916 and 1953, of patients with cerebellar signs attributed to parietal lesions. Extensive lesions within the parietal lobes and sensory changes were common in these patients. They also reported on an adult who presented with pain and weakness of the left arm, lasting for 2 hours. When examined 2 weeks later, the limb was severely ataxic, but with no detectable weakness or sensory deficit. In addition, the patient had a dressing apraxia. At autopsy a small infarct in the arm area of the right postcentral gyrus was observed. The cerebellum and brainstem were normal.

Ghika et al.[2] reported on an adult patient who had a history of recurrent transient ischaemic attacks, with recent pronounced clumsiness of the right hand, accompanied for a few minutes by tingling of the lower-left half of the face and a left temporal headache. The right arm had decreased motility and hypotonia, but no weakness. The finger–nose test indicated hypermetria, asynergia, and dysdiadochokinesia; and rhythmic movements of the right hand were grossly abnormal. All these findings were no worse with the eyes closed. Slow distal mobility, with an equivocal plantar response without ataxia, was the only abnormality of the right leg. Repetitive testing revealed no evidence of any impairment of proprioceptive sensation. The CT scan revealed a cortico–subcortical parietal lesion in the left cerebral hemisphere, and a left posterior temporal arachnoid cyst. Abnormalities in the parietal and temporal regions were later confirmed by MRI scan. Angiography showed a 70% stenosis of the right internal carotid artery at the carotid syphon. No lesion or atrophy could be seen in the cerebellum or in its pathways in the brainstem, or in the thalamic nuclei or elsewhere in the white matter.

These findings indicate that lesions in the posterior parietal area can mimic cerebellar ataxia, possibly by interrupting specific projections to the ventrolateral thalamic nuclei. Parietal ataxia is usually considered to result from a loss of proprioceptive feedback, interfering with the smooth execution of motor functions; and it is made worse by loss of vision. However, this was not the case in the patient Ghika et al.[2] described. Therefore, it appears that parietal ataxia can occur with no sensory loss.

Guard et al.[3] described an adult with bilateral parietal lesions, a glioma on the left side and a haematoma on the right, who presented with Balint syndrome (loss of automatic eye movements) and who also had ataxia predominantly of the right hand.

Steinlin et al.[4] reported on two boys, aged 3 and 4 years, with congenital cerebellar ataxia; investigations suggested a parietal origin. At an early age both boys had evidence of overall delayed development. When examined at the age of 3 years, one boy had hypotonia with mild ataxia on reaching, and walked on a broad base with truncal ataxia. His features were mildly dysmorphic. The second boy presented at 6 months of age with infantile spasms which responded to treatment. However, he has since had occasional seizures. He had dysmetria at 1½ years, and walked with a broad-based ataxic gait at 2½ years. At age 4 years he had significant hypotonia and ataxia. The EEG of this patient was firstly hypsarrhythmic, and later indicated focal abnormalities in the parietal areas. The MRI in both boys revealed marked parieto–occipital pachygyria, while the cerebellum was normal. The family histories of both children were not significant. Congenital ataxia is usually due to infratentorial abnormalities, but in these two subjects the ataxia appeared to be related to the bilateral parietal lesions. A disruption of cerebello–thalamo–parietal connections could explain these findings.

When reviewing non-progressive congenital ataxia, Steinlin et al.[5] reported that some patients had no cerebellar hypoplasia and, although associated symptoms such as epilepsy and cognitive disabilities suggested supratentorial involvement, there was no definite evidence of parietal lesions. However, in the study of non-progressive ataxia by Esscher et al.[6], a number of children had definite supratentorial abnormalities. Their motor disability was milder than those with infratentorial pathology. The lesions included cortical dysplasias and areas of abnormal migration, but localization to the parietal areas was not highlighted and some children had damage to various other areas of the brain.

Other examples of non-progressive congenital ataxia have been recorded. For example, this diagnosis was given in two sisters who both had extensive neuronal migration abnormalities[7]. Their development was delayed. The eldest, at the age of 9 years, had nystagmus, intention tremor on finger–nose testing, and an unsteady broad-based gait. The white matter of the parietal lobes had low density areas with CT scanning, and the MRI revealed dilatation of the ventricles and basal cisterns, and thickening and abnormal smoothness of the cerebral cortex, especially in the frontal and parietal lobes. The white matter was generally thin, with multiple regions of abnormal signals supratentorially, and

the cerebellar peduncles and cerebellar white matter were also affected to a lesser degree. The younger sister's disabilities were not so severe, but at age 5 years she had nystagmus, intention tremor, and an ataxic gait. Her CT and MRI scans showed similar changes to those of her sister. The EEGs of both children were characterized by unusual patterns of high amplitude, 10 to 12/s rhythms mainly anteriorly. It was considered that the findings were compatible with migration defects which might be due to an autosomal recessive disorder, but the abnormalities were too widespread to locate the responsible lesion.

Children with disorders of coordination as described could be classified under the rubric of ataxic cerebral palsy (CP). Miller and Cala[8] examined 29 patients with ataxic cerebral palsy, correlating the clinical findings with the CT scans. The CT scans were normal or only slightly abnormal in 38%, revealed posterior fossa lesions in 28%, and obvious cerebral abnormalities in 55%, which always involved the parietal lobes. All subjects with simple ataxia had abnormal CT scans, but the changes were widespread. The CT findings had a better correlation with IQ and epilepsy than with the clinical subtype. This study confirms that the diagnosis of ataxic CP is not synonymous with a cerebellar lesion, but may be due to an inability to integrate, at cortical and subcortical levels, the functions essential for posture, equilibrium, and coordination of movement.

The syndrome of parietal ataxic hemiparesis has various features[9]. The combination of ipsilateral corticospinal tract signs and ataxia usually suggests a lesion in the pons at the level of the junction of the upper one-third and lower two-thirds on the opposite side to the neurological signs; although, as reported in the article by Yagnik et al.[9], it has been associated with pathology in a number of sites, including the internal capsule, corona radiata, thalamus, and midbrain. These authors also reported on an adult patient who had weakness and ataxia of the left arm, and ataxia of the left leg. The CT scan revealed a well defined lesion in the right parietal area compatible with an infarct. Apart from the slight weakness, the subject had a drift of the left arm and a minimally impaired pain sensation of this limb with agraphesthesia and astereognosis. The finger–nose test revealed marked ataxia out of proportion to the weakness, which did not change when the eyes were open or closed.

Bansal and Chopra[10] reported on two patients, both of whom were 15 years of age, who presented with focal motor fits. The first patient had a 5-day history, followed by weakness and ataxia on the side of the seizures. The latter responded well to treatment, and the clinical signs resolved. The EEG was abnormal in the right centroparietal area. The second patient had a 2-month history of myoclonic jerking of the right hand, and pyramidal signs and ataxia affecting the right side. A CT scan showed a lesion in the temporoparietal area. After treatment with antiepileptic drugs, the abnormal signs resolved within a few days. The ataxic hemiplegia, after the epileptic fits, was presumably due to a functional deficit of the corticospinal and corticoponto–cerebellar fibres, and may have been due to circulatory or metabolic changes.

Conclusions

Ataxia is usually due to impaired function of the cerebellum or its connections, but can result from spinal-cord and peripheral nerve lesions. Ataxia associated with parietal lobe lesions has occasionally been attributed to loss of proprioceptive sensation, or to lack of spatial orientation; and it could be due to the destruction of the parietopontine fibres destined for the cerebellum. Sometimes the ataxia associated with a parietal lobe lesion can occur without any sensory loss. Apart from neuroradiological tests, other signs may help to localize the lesion. For example, the presence of nystagmus, dysarthria, and normal sensation indicates a pontine localization; and the presence of sensory abnormalities with the absence of dysarthria and nystagmus indicates a supratentorial localization and possible hemiparesis[9].

Ghika et al.[2] reported on a subject with ataxia without sensory loss and with a posterior parietal lesion, which was thought to be due to a lesion of the specific projection of the parietal lobe into the cerebellothalamic pathways, probably from area 5. Freund's studies indicated that lesions in the lateral parts of the premotor field interfered with proximal muscle function and interlimb coordination between both sides, although distal motor activity and bimanual coordination were unimpaired, so-called frontal lobe ataxia; whereas, in the presence of posterior parietal lesions, hand function was severely disturbed, especially purposive behaviour of the hand during exploratory finger movements and manipulation of objects[11]. The causes of this included ataxia, dysmetria, and postural instability, findings typical of deafferation. It was concluded that lesions of the frontal motor fields affected posture and force control; and parietal–cortical lesions, mainly in the posterior parts, were associated with a failure of sensorimotor integration and the elaboration of motor programmes that can generate purposive movements for exploring and manipulating objects with the hand. The parietal area is also involved in the control of visually guided movements[12].

In his studies on parietal lobe function, Critchley described proprioceptive ataxia with decomposition of movement, hypo- and hypermetria, intention tremor, hypotonia, and falling away of the outstretched hand on holding out the arm[13]. Also, rapidly alternating movements and the heel–knee test were poorly performed; Ghika et al.[2] have recorded such ataxia with a parietal lesion and no proprioceptive loss. Therefore, it is important to consider involvement of the parietal lobe as well as the cerebellum when evidence of ataxia is found.

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