Pediatr Radiol (1997) 27: 139–143

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Cerebellar infarction and atrophy in infants and children with a history of premature birth

Received: 24 November 1995
Accepted: 9 September 1996

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Abstract *Background and objective.* We wished to determine the pattern of cerebellar disease in children with a history of premature birth and early ultrasound evidence of intraventricular haemorrhage and/or parenchymal lesions of the cerebral hemispheres.
Materials and methods. MRI findings for all premature infants examined in a 3-year period (73 patients) were reviewed to determine the nature and frequency of lesions of the cerebellum and the results were correlated with clinical data.
Results. Six cases of unilateral cerebellar infarction were identified. These involved the posterior inferior cerebellar territory in each case (as well as other territories in two cases). A case of generalised cerebellar atrophy and three cases of unilateral cerebellar hemisphere atrophy were identified as well. In nine of these ten cases abnormalities were also seen elsewhere in the brain.
Conclusion. The literature describes cerebellar infarction in infants and children as rare, but this study shows that it is not unusual following perinatal haemorrhagic/ischaemic anoxic injury. It is suggested that cerebellar atrophy may also occur as a result of vascular disease.

Introduction

Cerebellar infarction is regarded as a rare condition in infants and children. The total number of cases described in the English-language literature in 1992 was only 33 [1]. The two largest published series both describe three cases [2, 3]. The aetiology was unknown in about half of the cases. In the other cases, trauma and physical exertion (six cases), congenital vascular and cervical spine anomalies (two cases), sepsis (two cases), dehydration (one case) and migraine (two cases) were implicated. We have retrospectively reviewed all MRI studies performed in infants and children with a history of premature birth at this institution (Hammersmith Hospital) over a 3-year period and identified all those with lesions involving the cerebellum.

Materials and methods

The MRI scans of all infants and children with a history of premature birth between September 1991 and August 1993 (73 patients) were examined and all those with cerebellar abnormalities were reviewed in detail. A detailed neurological assessment including evaluation of posture, tone, power and reflexes, with special attention to cerebellar function, was performed at the time of the MRI scan. All MRI examinations were performed on a 1.0-T HPQ scanner (Picker Ohio, USA). Sequences were: T1-weighted spin echo (SE; TR 620–920/TE 20), T1-weighted inversion recovery (IR; 2500–3500/30/TI 600–950) and T2-weighted SE (2500/20, 80, 120).

The MRI scans were reviewed by three observers. Infarction was diagnosed when a lesion with a well-defined boundary was present in the distribution of one of the cerebellar arteries while the remainder of the cerebellum was normal. Reference to published studies in adults was used to assign the lesion(s) to a specific cerebellar artery territory [4, 5]. Cerebellar atrophy or hypoplasia was diagnosed when one or both hemispheres or the vermis was decreased in size but the basic configuration of the cerebellum was preserved.

Table 1 Clinical and US results (*ELCS* elective low segment caesarian section, *GLH* germinal layer haemorrhage, *IVH* intraventricular haemorrhage, *L* left, *PHVD* posthaemorrhagic ventricular dilatation, *R* right, *SVD* spontaneous vaginal delivery)

Case	Gestational age (weeks)	Birth weight (g)	Pregnancy/delivery	Ultrasound	Development	Neurological outcome
1	24	670	SVD	Bilateral IVH, R porencephalic cyst	Motor delay	Ataxia (upper limb > lower), R hemiplegia
2	33	1190	ELCS	LIVH + ? porencephalic cyst, bilateral PHVD	Motor and visuo-perceptual delay	R hemiplegia, epilepsy
3	25	724	SVD	IVH, PHVD (shunt)	Mild global delay	Mild hypotonia, ataxia (upper > lower limb)
4	24	602	Infection, β-haemolytic streptococcus/ forceps	IVH	Mild global delay	Ataxia
5	30	1110	Hypertension, thrombocytopenia/ ELCS	Bilateral GLH, IVH	Mild delay	Mild hypotonia
6	28	650	Oligohydramnios/ ELCS	Thalamic lesions	Moderate global delay	Ataxia gait, ocular apraxia, Epilepsy
7	27	1230	ELCS	IVH and parenchymal PHVD (shunt) involvement	Severe global delay	Quadriplegia, optic nerve atrophy, epilepsy
8	23	630	Breech	Bilateral IVH	Moderate global delay	Increased tone in the legs, brisk reflexes
9	27	1180	SVD	IVH + porencephalic cyst, PHVD (shunt)	Motor and visual-perceptual delay	L hemiplegia, epilepsy
10	26	1028	SVD	IVH with parenchymal involvement	Mild motor delay	Clumsy child, motor difficulties

Results

Ten children showing involvement of the cerebellum were identified. The age of the patients at the time of MRI ranged from 7 months to 8 years 10 months. All ten were born at between 23 and 33 weeks, gestational age. Details of clinical history, neonatal ultrasound studies and follow-up are summarised in Tables 1 and 2. Six of the ten patients showed signs of cerebellar infarction. This was identified in the posterior inferior cerebellar artery (PICA) distribution with vermis involvement (medial and lateral) in four cases (Fig. 1) and without vermis involvement (lateral) in another one. One of these patients also had anterior CA involvement. The remaining patient only had evidence of inferior vermis infarction (medial PICA).

Four children showed cerebellar atrophy which was generalised in one case (Fig. 2) and unilateral in three (e.g., Fig. 3).

Discussion

Six cases of cerebellar infarction and four cases of cerebellar atrophy were diagnosed on MRI scans from a group of 73 children with a history of premature birth (incidence 8.2 %), several of whom had had evidence of parenchymal abnormality suggestive of a haemorrhagic and ischaemic aetiology. One of these patients showed isolated cerebellar involvement, but in all the others imaging abnormalities were found elsewhere in the brain. These predominantly involved the cerebral hemispheres and took the form of ventricular dilatation, porencephalic cysts or periventricular changes. In all but one case intraventricular haemorrhage had been present on neonatal ultrasound. None of the patients in our cohort showed acute onset of cerebellar signs. Clinical signs of cerebellar involvement were found in only four of the six patients with cerebellar infarction and in none of the four patients with cerebellar atrophy. They all showed developmental delay and abnormal neurological outcome.

Previously reported cases of cerebellar infarction generally describe acute presentation in patients without a

Table 2 MRI results (*l* lateral, *m* median, *PICA* posterior inferior cerebellar artery)

Case	Duration of clinical follow-up	Age (corrected) at time of MRI	Cerebellar hemisphere (and side L/R)	Vermis	Fourth ventricle	Brain stem	Cerebral hemisphere	Lateral ventricles	Imaging diagnoses
1	5 years	3 years 6 months	Infarction hemisphere L	Infarction inferior	Normal	Rotated	Porencephalic cyst (L); porencephalic cyst, small (R), periventricular	Dilated (L), slightly dilated (R)	i) Unilateral hemisphere infarct and vermis (PICA m, l) ii) Bilateral porencephalic cysts iii) Periventricular change (Fig. 1)
2	2 years	8 years 2 months 10 years 2 months	Infarction hemisphere R	Infarction inferior	Dilated L, open R	Mild wallerian degeneration	L fronto-parietal porencephalic cyst, large	Very dilated L, dilated R	i) Unilateral hemisphere infarct and vermis ii) Wallerian degeneration on left iii) Large R fronto-parietal infarct iv) Ventricular dilatation PICA (m, l)
3	3 years 10 months	2 years 8 months	Infarction and hemisphere medial R	Infarct inferior	Dilated on infarct side	Normal	Cyst, R frontal	Dilated, irregular, periventricular change, cyst	i) Unilateral hemisphere and vermis (PICA m, l) ii) R frontal cyst iii) Periventricular change iv) Ventricular dilatation
4	2 years 6 months	9 months	Infarction hemisphere small L	Infarct inferior	Small	Rotated	Mild periventricular change	R ventricular, dilated, L slight	i) Small left infarct (PICA m, l) ii) R ventricular, dilated
5	1 year 6 months	7 months	Infarction R, cerebellar hemisphere R	Normal	Normal	Normal	Small infarction L, parieto-frontal	L ventricular, dilated	i) Unilateral hemisphere infarction (PICA m) ii) Fronto-parietal infarct iii) Mild ventricular dilatation
6	6 years	5 years 8 months	Inferior vermis infarction	Normal	Normal	Normal	Normal	Normal	i) vermis infarction (m)
7	3 years	3 years	Severe atrophy L and R	Severe atrophy superior and inferior	Grossly dilated	Moderate atrophy	Gross atrophy L and R, hygromas	Irregular, enlarged, L and R shunt	i) General atrophy, enlarged fourth ventricle ii) Brain stem atrophy iii) Hemisphere atrophy, wallerian degeneration iv) Bilateral hygroma (Fig. 2)
8	3 years	1 year 11 months	Moderate atrophy L	Normal	Normal	Normal	Normal	Moderate enlargement L and R	i) Unilateral cerebellar hemiatrophy ii) Moderate lateral ventricular enlargement
9	10 years	8 years 10 months	Moderate atrophy L	Normal	Small on left	Rotated atrophic	R severe atrophy, L slight atrophy	R distorted, L mild distortion, shunt	i) Unilateral cerebellar hemiatrophy ii) Severe atrophy R cerebral hemiplegia iii) Wallerian degeneration iv) Shunt
10	8 years 3 months	7 years 5 months	Moderate atrophy R	Normal	Normal	Normal	Periventricular change	R moderately dilated, L mildly dilated	i) Unilateral cerebellar hemiatrophy ii) Periventricular change iii) Dilated lateral ventricles (Fig. 3)

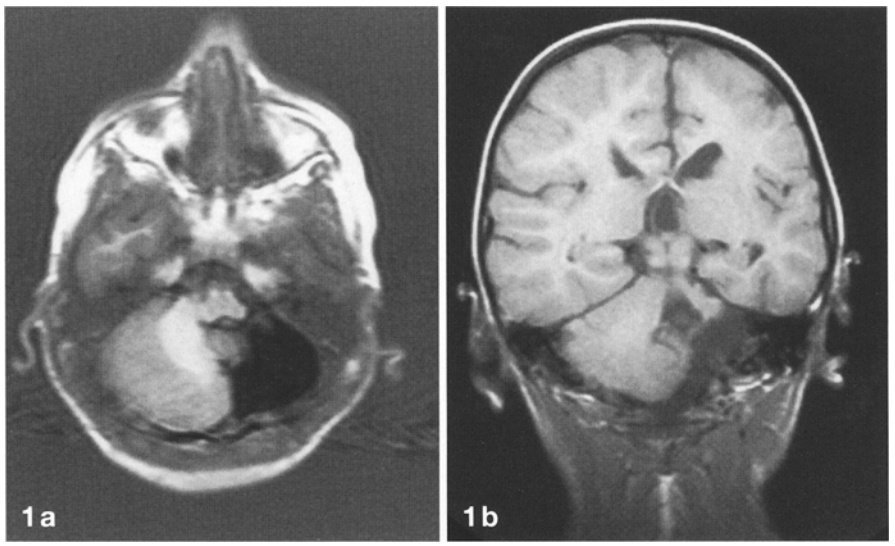
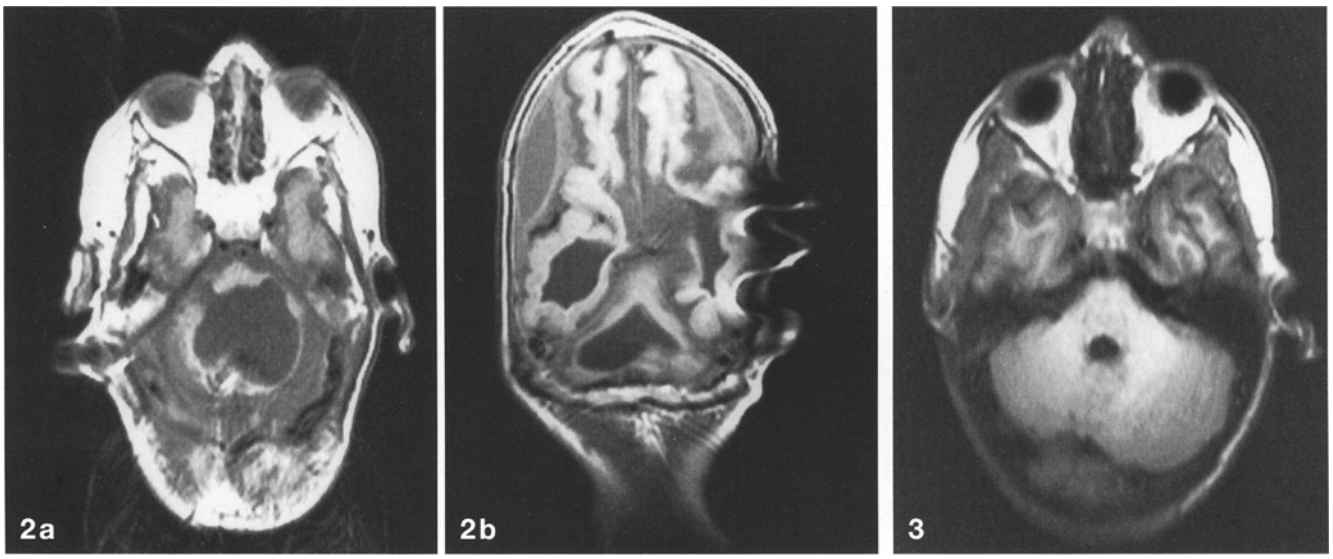


Fig. 1 a, b Infarction in the posterior inferior cerebellar artery territory (case 1): **a** Transverse IR TR 3100/TE 30/TI 700 and **b** coronal SE 660/20 images. The inferior left cerebellar hemisphere and vermis are absent and replaced by fluid

Fig. 2 a, b Generalised cerebellar atrophy (case 7): **a** Transverse SE 660/20 and **b** coronal SE 860/20 images. The fourth ventricle is dilated and the cerebellum is generally atrophic. The atrophic cerebral hemispheres, hygromas and shunt artifacts are noted

Fig. 3 Cerebellar hemisphere atrophy (case 10): Transverse IR 3100/30/700 image. The right cerebellar hemisphere is atrophic



previous history of ischaemic or haemorrhagic disease. A wide range of possible causes are described in these patients, including congenital heart disease, vascular malformations, trauma, infection, migraine, atherosclerosis, hypertension, collagen vascular disorders, metabolic disorders, haemorrhagic disorders, aminoacidurias, hyperlipidaemias, vasculitis, fibromuscular dysplasia and neurocutaneous syndromes [6]. The difference between our findings and those of previous authors reflects the facts that the study group reported here is a relatively new population in the historical sense, that the cerebellar lesions were frequently associated with disease elsewhere in the brain (and therefore may not have been specifically recognised) and that new and more accurate imaging techniques make diagnosis easier.

Cerebellar atrophy also has a wide variety of causes and associations [7], but the very similar histories of the two groups of infants in this series (i.e. those with cerebellar infarction and those with cerebellar atrophy) suggest that vascular events may account for the atrophic change. The unilateral occurrence in three cases and the occurrence of vascular changes elsewhere in the brain also support this hypothesis. It is possible that the atrophic pattern may have resulted from hypoxic ischaemic injury occurring in a form of the circulation more immature than in older infants, with a more general effect on the cerebellum or cerebellar hemisphere. There is one report of the pathological features of cerebellar atrophy in which perinatal vascular disease is emphasized [8]. The authors of this study drew attention to cerebellar sclerosis, in which atrophy and scarring of the cerebellum are combined. The sclerosis typically occurred either in the distribution of cerebellar arteries (or a border zone territory), indicating an ischaemic

cause, or, when diffuse, in the gyral depths, suggesting a hypoxic cause. They suggested that perinatal hypoxic ischaemic injury is the most important cause of non-progressive sclerotic atrophy of the cerebellum of this type.

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