Dysplastic gangliocytoma of cerebellum in a newborn

Diagnosis by ultrasonography and MRI

M. Dietlein, R. Schröder, B. Widemann, and G. Benz-Bohm

1 Department of Radiology, Pediatric Radiology, 2 Department of Pathology and
3 Children's Hospital, University of Cologne, Cologne, FRG

Received: 2 December 1991; accepted: 28 January 1992

Abstract. A cerebellar lesion was detected by ultrasonography (US) in a symptomatic newborn. Magnetic Resonance Imaging (MRI) demonstrated this lesion as a gyri-form structure. Therefore a rare dysplastic gangliocytoma was suspected, which was compatible with histopathology. Immunohistochemical studies (monoclonal antibody Ki-67) showed signs of progression.

Dysplastic gangliocytoma (Lhermitte-Duclos' disease, purkinjeoma, ganglioneuroma, hamartoma of the cerebellum, granule cell hypertrophy or granulomolecular hypertrophy of the cerebellum) is a rare lesion of the cerebellum, characterized by thickened and dysplastic gyri. Since the first description in 1920 about 75 cases have been published [1-17]. Computed Tomography (CT) and angiography are insensitive in imaging this specific tumor. Up to now the diagnosis was therefore established by histopathological examination, whereas the typical MRI pattern of this lesion leads to a preoperative diagnosis [2, 3]. In newborns and infants this mass is usually detected by US.

Case report

G.V., a full term newborn, was referred to the hospital on its third day of life because of episodes of central apnoea, bradycardia, nystagmus and deviation of the eyes to the left.

Family history: short stature of the 16 year old primigravida (150 cm), connatal dysmelia of her right hand.

Clinical findings on admission: laboured, irregular, shallow breathing, strabismus diergens intermittenst, nystagmus, oral automatics. Birth weight 3160 g, body length 48 cm, head circumference 35.5 cm. No malformations.

Sleep-electroencephalography: Multifocal spikes. After two seizures a therapy with Phenobarbital was initiated.

US of the head (3.5 and 5 MHz sectorscanner): median sagittal section: round, hypoechoic cerebellar lesion of 2.3 × 2.3 cm, reaching to the fourth ventricle. No hydrocephalus (Fig. 1).

CT: ill-defined 3 × 2.5 cm hypodense lesion extending into both cerebellar hemispheres with displacement of the fourth ventricle to the left and ventrally. No enhancement, no calcifications (Fig. 2).

MRI (SE; TR 500, 2300; TE 20,100; native, i.v. Gadolinium (Gd-DTPA) T1: the lesion is hypo-intense with respect to the white matter, measuring 2.5 cm in diameter and lying medially in the right cerebellar hemisphere. There is a pronounced gyriform appearance. There was no enhancement after Gd-DTPA. T2: increased signal intensity of the lesion similar to the cerebral cortex (Fig. 3b). Displacement of the fourth ventricle to the left and ventrally, impression of the brain stem (Fig. 3a). The pattern of pronounced gyri

Fig. 1. US of the brain (3.5 MHz), median sagittal section: round hypoechoic cerebellar lesion, 2.3 cm Ø, separation from the fourth ventricle impossible, relation to brain stem unclear. Newborn fourth day of life

Fig. 2. CT with contrast medium: no enhancement of the ill-defined hypodense cerebellar lesion, displacement of the fourth ventricle, relation to brain stem unclear

Fig. 3a,b. MRI. a SE 500/20, median sagittal section: cerebellar lesion with decreased signal, displacement of the fourth ventricle, impression of the brain stem. b SE 2300/100, transverse section: gyral pattern of the cerebellar lesion with signal intensity corresponding to cerebral cortex
No postoperative complications. In spite of continued anticonvulsive therapy cerebral seizures and facial automatons still occurred.

**Discussion**

Dysplastic gangliocytoma is caused by a severe derangement of the laminar architecture of the cerebellar cortex with features of a malformation and a neoplasm. In the transition area of normal to pathological cortex a gradual loss of Purkinje-cells, a hypertrophy of stratum granulosum and accumulation of abnormal shaped ganglion cells with myelinated axons in the molecular layer are observed [4-8]. Macroscopically there is a focal or diffuse thickening and paleness of the involved cerebellar gyri. The pathogenesis is unknown. A congenital disorder in the migration and development of the granule cells of stratum granulosum is postulated [8].

In our case the full histopathological picture could not be demonstrated because of the small specimen surgically removed. The partly negative immunohistochemical reaction to NSE must be seen against the background of the still immature central nervous system in this newborn. However, differential diagnostically no other known cerebellar lesion type than dysplastic gangliocytoma comes into question.

The growth fraction, demonstrated in this case, as well as the rare observation of mitoses and postoperative recurrences after long intervals are signs of sometimes progressive growth [7, 9].

The growth fraction of 0.4% is comparable with that of a slowly growing pilocytic astrocytoma [18].

Up to now dysplastic gangliocytoma was diagnosed in only one newborn at autopsy [10]. It was diagnosed in three patients during the first decade of life, mostly during the third and fourth decade [11-14]. Males and females are equally affected [14].

The clinical symptoms are caused by increased intracranial pressure. Only one fifth of the patients exhibit cerebellar signs [14-16]. The episodes of apnoea and bradycardia in our patient were signs of brain stem compression due to the cerebellar tumor.

Up to now in patients with gangliocytoma seizures only occurred with supratentorial tumors, as a consequence of cardiac arrest or in the early postoperative phase [10, 15, 19]. The cause of the seizures in our patient is therefore uncertain. Macrocephalus, hydrocephalus and hydromyelia are often associated with dysplastic gangliocytoma, whereas polydactyly, hemihypertrophy, macroglossia, mental retardation, neurofibromatosis, multiple hemangiomias and spongioblastoma were rarely observed [3-6, 8, 10, 15]. In spite of the occurrence in one family—a son and his mother—an inheritance pattern has not been identified [15].

In infancy US and MRI are the best imaging modalities. US serves as a screening investigation for early diagnosis. MRI is superior due to the multiplanar imaging capability in detecting cerebellar lesions and defining the size and relations of the fourth ventricle and brain stem (Fig. 3a). The typical gyral pattern demonstrates the cortical nature of this lesion [2, 3, 7, 16, 17]. Technical improve-