Acta Paediatrica Hungarica 30 (1), pp 43-51 (1990)

NEONATAL PERIVENTRICULAR LEUKOMALACIA: DIAGNOSIS AND MONITORING BY REAL-TIME ULTRASOUND

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Received 23 March 1988

Periventricular leukomalacia is a form of hypoxic-ischaemic encephalopathy developing in preterm babies. During the last year among 387 sonographically screened neonates 11 periventricular neonatal leukomalacia cases were found. Analysing their cases authors discuss its sonographic diagnosis and follow up.
In the acute stage characteristic triangular shaped area of increased echogenicity appears at the external angle of the lateral ventricles, later regular shaped echo-free areas of cysts appear in that region. The diagnostic and prognostic significance of cranial sonography in periventricular leukomalacia is discussed.

INTRODUCTION

Periventricular leukomalacia (PVL) is well recognized as a relatively uncommon, yet especially serious complication of prematurity. It represents an infarction of the periventricular white matter adjacent to the external angle of the lateral ventricles. Its hypoxic-ischaemic nature was documented by Banker and Larroche /1/. Later DeReuck et al /3/ suggested that this lesion is the anatomical basis of spastic diplegia.

Recent advances in imaging techniques have provided an opportunity for establishing the diagnosis of PVL in life, and following its evolution in relation to the clinical picture /4,6,7,9,10,11/.

Between the first of November 1986 and the thirty first of October 1987 we diagnosed and followed with ultrasound 11 cases of periventricular cystic leukomalacia. Our experiences are discussed in the followings.

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MATERIALS AND METHODS

Cranial ultrasound was performed on 387 neonates, using a Picker LS 7000 scan and a 5MHz convex transducer /20/. Eleven neonates were diagnosed as having PVL. All but one were of 32 weeks or less gestational age, and weighed less than 1800 g at birth. The neonates underwent routine cranial ultrasound examination during the first week of life, and again at about 3-4 weeks of age. If either sonogram showed any evidence of intracranial abnormality the scanning was continued, every one or two months subsequently. Two of the PVL babies ("outpatient babies") had only "late" sonograms.

Neurological examination was carried out in the first week of life, then three-monthly, up to one year of life, in all cases.

PVL was diagnosed during the first week of life if a triangular area of increased echogenicity appeared at the external angle of the lateral ventricles (Fig. 1). The development of anechoic regularly-shaped areas (cysts) in that area confirmed the diagnosis (Fig. 2 and 3).

All but the two "outpatient babies" had neurological assessments in the newborn period, and then on a three-monthly basis and at one year of age.

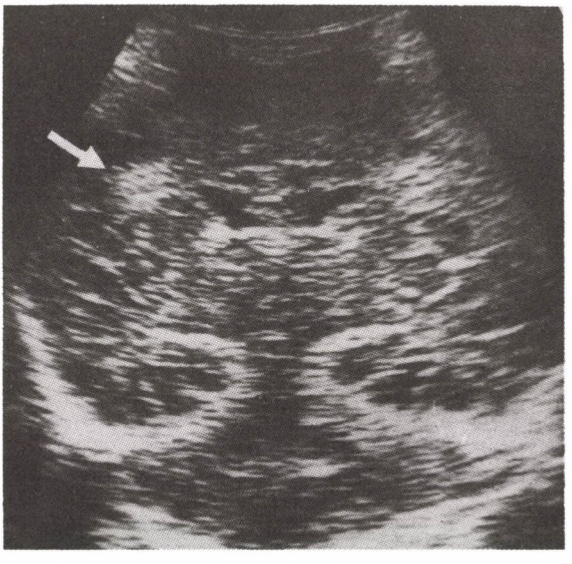

Fig. 1. Triangular shaped increased echogenicity at the external angle of the lateral ventricles

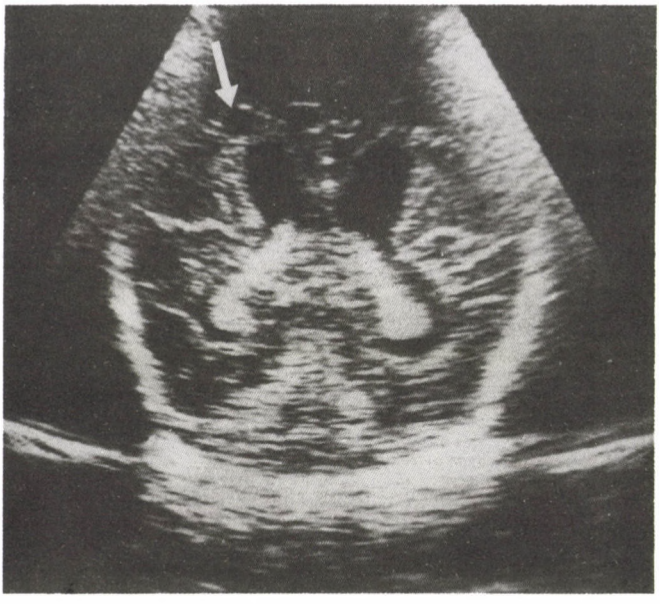

Fig. 2. Periventricular cyst on posterior coronal section

RESULTS

Among 387 neonates eleven showed the sonographic signs of periventricular leukomalacia. In two cases significant periventricular haemorrhage appeared in addition to PVL.

Periventricular cysts developed by the third or fourth week of life. In one case the increase in periventricular echogenicity persisted for three weeks but without cystic formation (Fig. 1).

In two patients the periventricular cysts involuted in two to three months, with mild enlargement of the lateral ventricles. In one child septations between the periventricular cysts disappeared in three months and later degeneration of septa and ependyma resulted in formation of a large common cavity (Fig. 4).

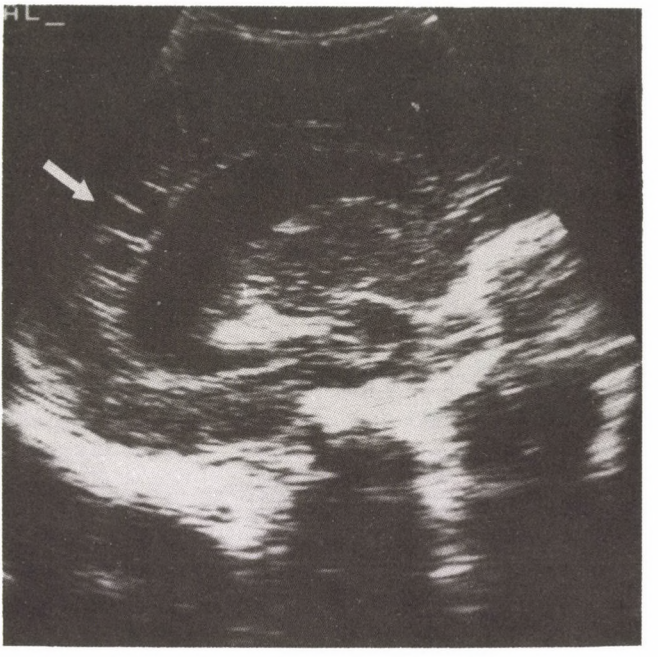

Fig. 3. Cysts on parasagittal section

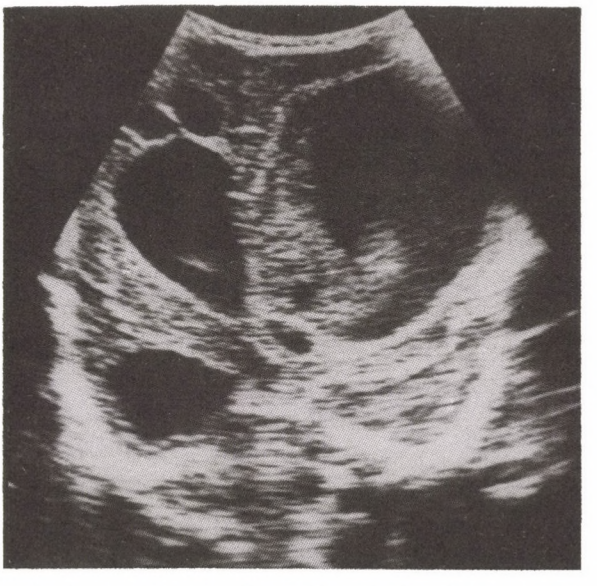

Fig. 4. Ventriculomegaly and periventricular cysts on the left side, a large common cavity "pseudoventricle" on the right side (coronal section)

All but one baby is now over one year old. One baby is hemiplegic (with contralateral periventricular cysts), another has spastic diplegia, the others are all tetraspastic or tetraplegic. Three have visual impairement (all with posterior-located cysts).

DISCUSSION

Periventricular leukomalacia of the cerebral white matter occurs at a characteristic site in the suppralateral angle of the lateral ventricles. This is the arterial boundary zone between the ventriculofugal and ventriculopedal arterial circulations (Fig. 1). In addition to inadequate anastomoses, poor development of the ventriculopedal arteries in the preterm infant may contribute to the specific vulnerability of this brain area /13,19/.

A significant history of birth asphyxia is common among the PVL patients. The vulnerable areas are particularly at risk during perinatal hypoxic episodes, when the cerebral autoregulation is impaired /12,14/. Hypercapnia, acidosis and hypoxia are all signficantly associated with the development of PVL /12/.

Although the aetiology of PVL may be border-zone infarction, more than one aetiological factor is probably involved. Pape and Wigglesworth /13/ described congestion of the branches of the terminal vein, in cases of massive germinal matrix haemorrhage with associated venous infarction of the surrounding periventricular white matter. This would account for the incidence of PVL in infants with significant (grade II-III) haemorrhage.

There are no early neurologic findings specific for PVL. Seizures, apnoeic attacks, disturbances in consciousness, abnormal muscle tone and leg weakness may all be found /21/.

Following the hypoxic-ischaemic events coagulation necrosis occurs in the periventricular white matter, followed by astrocytic, endothelial and macrophage proliferation.

Phagocytosis of the necrotic tissues results in periventricular cavities surrounded by gliosis. The thinning of periventricular white matter can lead to ventriculomegaly /4,9,21/.

The development of PVL can be followed by real-time ultrasonography. In the acute stage characteristic triangular-shaped areas of increased echogenicity are present at the external angle of the lateral ventricles seen in middle coronal sections of the brain. As the infarctions are frequently bilateral the areas of increased echogenicity are also usually bilateral and symmetrical. Although echogeneity usually appears internally homogenous, scattered areas of higher and lower level density may sometimes be seen within. The lateral contour is often irregular /16/.

At times it may be difficult to differentiate PVL from a normal periventricular echogenic halo. The development of periventricular cysts would confirm the original diagnosis /16/. Diagnosis is more difficult when PVL and periventricular haemorrhage occur simultaneously. Differentiation between the two is very important, as PVL - unlike haemorrhage - carries a universally poor prognosis (spastic diplegia, quadriplegia, visual defects, developmental delay). Subependymal haemorrhage may extend into the periventricular white matter. The characteristic positioning of PVL, dorsal and lateral to the germinal matrix, helps to differentiate these entities /2,8/.

At the time of intense phagocytosis the brain can appear sonographically normal. Three to four weeks after a hypoxic-ischaemic insult the periventricular infarctions undergo liquefaction, and regular echofree (cystic) areas appear. The cysts can be small (less than 3 mm in diameter), or larger, they may develop anteriorly or posteriorly to the trigonum, or follow the entire contour of the lateral ventricles. Septa between the cysts may degenerate, as may eventually the ependyma lining the lateral ventricles. This constitutes development of "pseudoventricles" (Fig. 4): unlike hydrocephalus, this situation responds poorly to shunting.

One of our sequentially-scanned babies had a persistent hyperechoic appearance of PVL without cyst formation. He is now

one year old, and tetraspastic. Some authors suggest that persisting increased echogenicity is likely to reflect permanent microscopic changes in the parenchyma: spongiosis microcalcification /3,17,18/.

The well known sequelae of PVL are spastic diplegia and quadriplegia, following corticospinal tract impairment at the external angle of the lateral ventricles, and visual and hearing impairment, when the periventricular cysts are posteriorly located.

Recognition of PVL is of considerable clinical significance. It implies significant brain damage and an unfavourable outcome. Establishing a correct diagnosis may influence early treatment (particularly with respect to long-term ventilation and perfusion) and later management, institution of rehabilitation and family counselling /2/.

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