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The Role of CT-Imaging in Recognizing the Type of Hydrocephalus and Porencephaly in Children

Rola TK w rozpoznawaniu typu wodogłowia i dziurowatości mózgu u dzieci

Intrauterine USG examination becomes more and more important in diagnosing developmental lesions of cerebral nervous tissue. However, CT--imaging is a method of choice, especially after parietal closure (13, 16). The character and etiology of porencephaly, congenital or acquired, is diversiform and often difficult to recognize (18).

This paper presents an attempt at assessing different forms of porencephaly, taking into account congenital hydrocephalus in children by means of CT--imaging.

MATERIAL AND TECHNIQUE OF EXAMINATION

The material comprises a group of 32 persons of both sexes, aged from 1 month to 12 years with congenital hydrocephalus. They were hospitalised in the District Hospital of Kielce in the years 1990—1993. CT examinations were performed with generation IV equipment of high definition produced by Siemens, Somaton H and Q type with 512×512 matrix. Sections were performed following the generally accepted norm (11), as a rule 8.4 or 2 mm thick. In patients with pathologic changes multiple CT examinations were performed after injection IV of 75% Uropoline in the bolus form in the amount of 2 ml/kg body weight. There were also done reconstructions in the sagittal and frontal surface.

CT examination in infants and small children often involved the necessity of short-term general anaesthesia. Thiopental in the amount of 2—4 mg/kg body weight. The examination technique of small children was according to the descriptions in the literature (11).

RESULTS

Hydrocephalus was accompanied by liquid cavernous spaces within the cerebral nervous tissue (porencephaly — 23 cases, arachnoidal cysts — 5 cases, Dandy-Walker's syndrome — 2 cases, hydranencephalus — 2 cases).

Standard X-ray pictures of the skull showed X-ray features characteristic of increased intracranial pressure, while clinical examination revealed an excessive increase of head circumference and fontanelle tension. Cavernous structures occurred: single in 13 and multiple in 19 children. In 9 children cavities with homogeneous fluid density showed communication with widened ventricular system of internal and external porencephaly type (Fig. 1). In 3 children there was shown a big cavity communicating with the extended arachnoidal space of external communication type (Fig. 2). In 2 children single cavities were classified as forms of closed porencephaly. They did not show increased density after interavenous administration of contrast medium. Their walls did not reveal any trace of circinate strengthening occurring in cystic changes of a different etiology.

Measurements of the cavities densities let differentiate brain mantle from porencephaly (15). Density of 15—25 CT number was shown by areas of morbid softening resulting from ischaemia or congenital postinfectious changes while the fluid in the cavities and porencephaly showed the density of 3—15 CT number.

In 3 children with porencephaly communicating with the ventricular system a wide, atrophic arachnoidal space was visible. In 2 persons big hemispheric cavernous structures were regarded as hydrocephalus.

Congenital changes were in two cases diagnosed in the form of Dandy-Walker's syndrome (Fig. 3). In 1 child congenital changes consisted in a partial defect of the vermiform lobe and hypoplasia of cerebellar hemispheres.

In 2 cases arachnoidal cysts were localized in the posterior cavity (Fig. 4) and in 2 cases on the fornix in the temporofrontal region.

DISCUSSION

Early brain injuries occurring in the first half of pregnancy, called pseudo-porencephaly, cause growth disturbances of the brain mantle. They result from a congenital infection, vascular occlusion and subsequent infarction. Early damages cause formation of cavities without any glial reaction in the surrounding region because of total resorption of necrotic cerebral tissue and these often coexist with congenital hydrocephalus (13). Similarly, porencephaly can result from perinatal damages leading to circulatory disturbances, intracerebral haemorrhages, infarction or result from a past infection in the last trimester of pregnancy (14). Kesaree's experiments (6) performed on animals indicate the

mechanism of porencephaly formation because of postoedematous necrotic area resulting from intrauterine ischaemia (5, 6). Porencephaly originating in the IInd half of pregnancy because of colliquative necrosis indicates thin-walled liquid cavities and does not communicate with the ventricular cerebral system (15). The temporal region is especially prone to the formation of colliquative necrosis and origination of porencephaly (7). The brain damages under discussion occur more frequently in prematurely born children (12). They are subsequent to subependymal bleedings also leading to communicating hydrocephalus. An immature, non-myelinized brain is more susceptible to ischaemia.

Periventricular encephalomalacia of the white matter in newborns also leads to the formation of cavernous structures. They are differentiated with incomplete myelinization which is accounted for by lack of periventricular oedema in the control CT examination.

After birth the brain responds to all injuries by glial fibrosis with only partial resorption of necrotic tissue. Fibro-glial cavities are formed (multicavernous softening). The cavity has partitions with an irregullar glial wall. Numerous cavities in the necrotic area are separated by the glial tissue. The glial reaction lets differentiate congenital defects of the immature brain from postnatal destructions (15).

Damages of the outer parts of the hemispheres with preserved perinatal space account for an acute episode of increased intracranial pressure of ischaemic character. The walls of extended ventricles are then separated from the fornix by the gradually necrotising brain mantle. A sudden increase of intracranial pressure may encumber peripheral circulation on the fornix, while central tissues are buffered by ventricular spaces. The ventricles can also extend secondarily when ischaemic tissue does not resist the pressure of the ventricular fluid.

Hydrocephalus of a considerable degree is differentiated from hydranencephalus, bilateral porencephaly, massive congenital subscleral hydrocele, incomplete division of the forebrain, postinfectious encephalopathy or with ischaemia.

A massive congenital subscleral hydrocele forms the central wedge-shaped reservoir of fluid, or tent-like configuration of medial perisicklar structures.

Hydranencephalus (vesical brain) and the extreme form of bilateral porencephaly are thought to have a common vascular etiology (5). No ependymal cells are observed on the internal surface of the membranous vesicle as a result of intrauterine infarction of the hemispheres. Different degrees of aqueduct narrowing account for the presence of the hydrocephalic component in hydranencephalus (5). There is emphasized a sudden, excessive growth of the infant's head circumference occurring after birth in the first weeks of life. According to Billewicz (2) cerebral angiography has the crucial role in diagnosing the disease.

In the cases of Dandy-Walker's syndrome there was shown a remarkable widening of ventricle IV with coexisting hypoplasia of the cerebellar vermiform

lobe and in 1 case it did not exist at all. The hypoplasia also involved the cerebellar hemispheres. The degree of ventricle IV widening ranged from the diverticulum extending back between the hemispheres in the site of the hypoplastic inferior vermiform lobe to the cystic formation involving the widened posterior fossa. As a rule, the cover of ventricle IV is formed of thin ependymo-arachnoidal membrane, the pons is sometimes displaced forward, depressed and deformed, and the reservoirs are constricted (18). The coexisting abnormalities in the form of narrowed aqueduct, lack of corpus collosum and transparent septum (interhemispheric cyst) or with the retention of the septum encountered in 68% of Dandy-Walker's syndrome (8, 9). The syndrome should be differentiated with the arachnoidal cyst, isolated ventricle IV, giant cerebello--spinal reservoir and porencephaly (1). Widenings of ventricle IV can also result from inflammatory-adhesive changes of the ependyma in the fetal period. The isolated ventricle is then of spheric shape while the present cerebellar vermiform lobe does not show abnormal structure. The arachnoidal cyst, pushing cerebellar hemispheres forwards, forms a liquid space at the back of the cerebellum.

The extensive cerebello-spinal reservoir can resemble an arachnoidal cyst. Yet, it does not dislocate the structures of the posterior cranial cavity.

Arachnoidal cysts recognized in 5 children result from the disorganization of the fetal arachnoidea or intrauterine bleeding (3). They are thought to be usually localized in the posterior cranial cavity (42%) and in the anterio-inferior region of the temporal lobe (18). In the combination with the lobe's hypoplasia they give a minimal dislocation of cerebral structures. On the cerebral fornix they have the shape of bi-convex lens with liquid density that does not change after the administration of contrast medium. M e n e z e s et al. (10) think that the cyst often coexists with other abnormalities (20—30% of cases). There is observed the coexistence of hydrocephalus, arachnoidal cyst and porencephaly. The above mentioned cavernous structures can get wider in the way of the valvular mechanism (4, 17) while hydroceles are of a slightly different shape because they are more shallow and extensive.

Conclusions

1. CT examination in the course of infantile hydrocephalus can show liquid cavities of the brain and the kind of their communication.

2. Diagnosing the character of communication of cerebral liquid spaces enables decompressing treatment (implantation of a ventriculo-peritoneal valve).

3. In order to establish the etiology of the cavernous structures accessory examinations should sometimes be performed (angiography, perfontanellar ventriculography, computing tomocysternography, infusive tests).

4. Final diagnosis of porencephaly is made by control CT examinations when inital examinations reveal multiple infarction areas of the brain.

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EXPLANATION TO FIGURES

Fig. 1. Male patient Z. M. aged 18 months. Porencephaly of internal and external communication type and of big hydrocephalus.

Fig. 2. Male patient. F. E. aged 8 months. A big cyst in the left temporal lobe, widely communicating with the subarachnoidal space and with atrophic widening of the left lateral ventricle.

Fig. 3. Female patient S. K. aged 4 years. Congenital hydrocephalus. Pudenz's valve was implanted at the age of 2 weeks, removed in the 12th month of life and implanted again 3 months ago (epileptic attacks). Lateral ventricles form an interhemispheric liquid cavity (lack of the great cerebral commissure). An extensive cystic space in the medial surface of the posterior cranial cavity (lack of cerebellar vermiform lobe and hypoplasia of the hemispheres account for Dandy-Walker's syndrome).

Fig. 4. Female patient M. M. aged 3 months. There was shown a big arachnoidal cyst in the posterior cerebral cavity with clear contours. Its density equalled the density of the cerebro-spinal fluid. After administration of Uropolin no contrast intensification was shown (confirmed operatively).

STRESZCZENIE

Na podstawie piśmiennictwa i badań TK u 32 dzieci obojga płci w wieku od 1 miesiąca do 12 lat podjęto próbę oceny rozpoznawania różnych typów dziurowatości mózgu z uwzględnieniem wrodzonego wodogłowia komunikującego. Rozpoznanie charakteru komunikacji przestrzeni płynowych i dziurowatości mózgu daje podstawę do leczenia odbarczającego i przewidywania jego skuteczności.

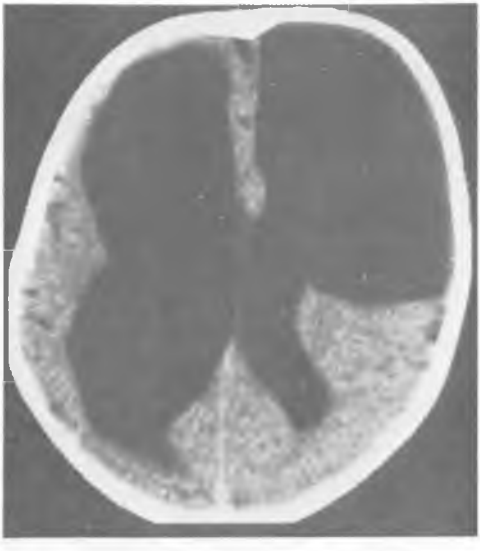

Fig. 1

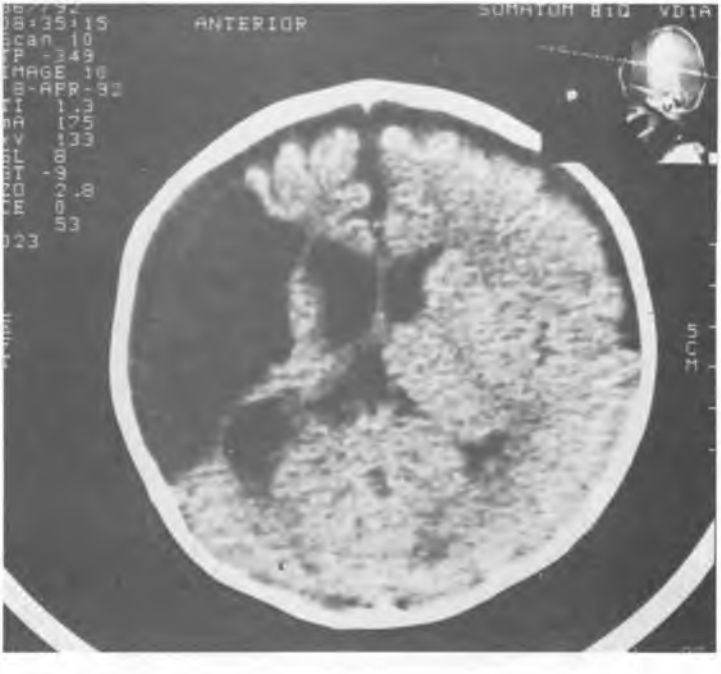

Fig. 2

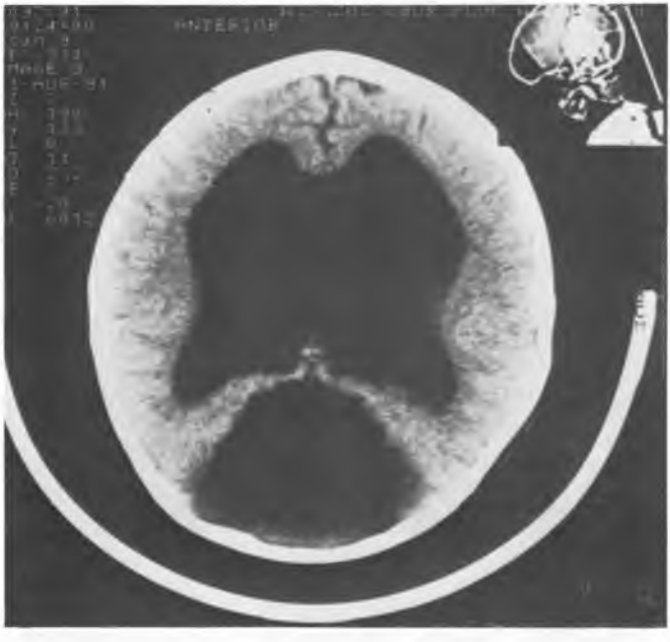

Fig. 3

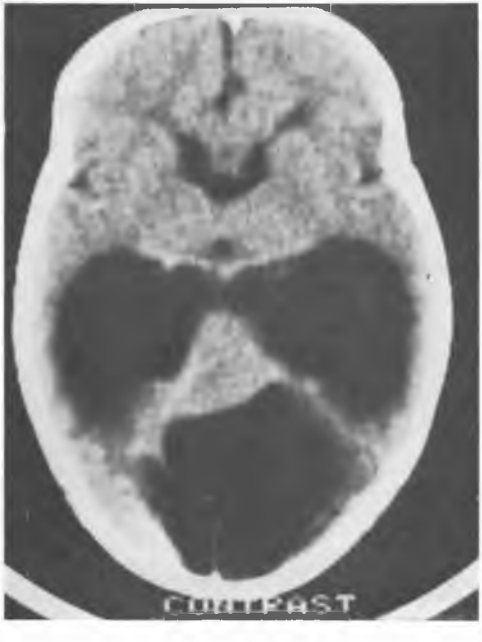

Fig. 4