I. NEONATOLOGY

A FORM OF DANDY-WALKER SYNDROME AS A PART OF A MULTIPLE MALFORMATIVE SYNDROME. LIMITATIONS OF THE ULTRASOUND SCAN.

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Abstract

The most impressive issue of this special case is that altrough standard ultrasound scan correctly recommended and performed during the pregnancy do not always identify some of the cerebral malformations. When dealing with this specific type of malformations, they can "escape" the prenatal ultrasound screening and therefore they get diagnosed postnataly.

In this particular case the hydrocephalus was identified not only through a clinical examination or ultrasound scan but mostly by magnetic resonance.

Key-words: Dandy-Walker Syndrome, ultrasound scan.

History

First carrying 32 years old woman who followed precisely the doctor's recommendations regarding medical examinations, clinical follow-up and ultrasound scans during the pregnancy; the risk score for the 21st trisomy ranged 1.1674 (embriofetal biometry, free serological βHCG and αFP).

The ultrasound scan performed at 20 weeks of gestation did not identify any foetal malformations, the intracranial structures appeared normal, the cerebellum's vermix was obvious (while using the standard incidence), the cisterna magna seized 6 mm, normal figure of the ventricles.(fig.1)

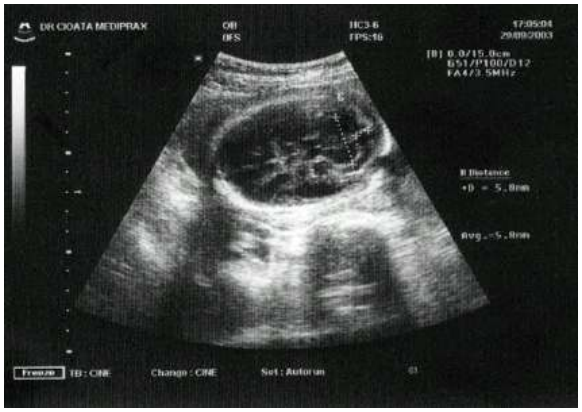

Fig. 1. Ultrasound scan at 20 weeks of gestation – normal aspect of the ventricles.

At 31 weeks of gestation there is noticed a premature imminence of the delivery. That was successfully stopped using tocolisis, uterosedatives and antispastics drugs. Accordingly, it was decided to adminster corticosteroids in order to prevent the hyaline membrane disease (Celestone VI f). On the ultrasound scan there was evident a bordered placenta praevia.

Birth: at 33 weeks of gestation a minor bleeding episode intervened being shortly after followed by a massive haemorrhage. The caesarean section was the best choice of that moment and so it was performed as an emergency. There it was, a new born baby girl, weighing 2140 grams, Apgar scored 6/7 at 5 minutes who was in obvious need of bag-and-mask ventilation using 100% O_2 than 40%O_2. The obstetrician confirmed his suspicion of accrete bordered placenta praevia.

The new born baby was admitted in the Neonatal Intensive Care Unit undergoing continuous monitoring. The first chest X-ray performed soon after birth showed a fixed lifting of the right hemi diaphragm which rose the question

of a possible congenital paresis of the right frenic nerve. The peribronchovascular image sustained a lack of amniotic fluid resorbtion (considered at that time as a neonatal transient tachypnea). The baby's condition aged 24 hours was worsening. She was experiencing a moderate respiratory distress syndrome (scoring 4/5 on Silverman scale) and so the neonatologist decided to intubate her for starting conventional mechanical ventilation. The chest X-ray performed in this stage undoubtfully showed a right pneumothorax and pneumomediastine (fig.2,3,4). The symptoms could be kept under control while receiving the mechanical ventilation via the endotracheal tube and while performing the aspiration puncture on the right hemi diaphragm. But not for so long and there was a need for a second puncture. At 4 days of age, undergoing conventional mechanical ventilation and pleural drainage, there were no signs of respiratory distress whatsoever.

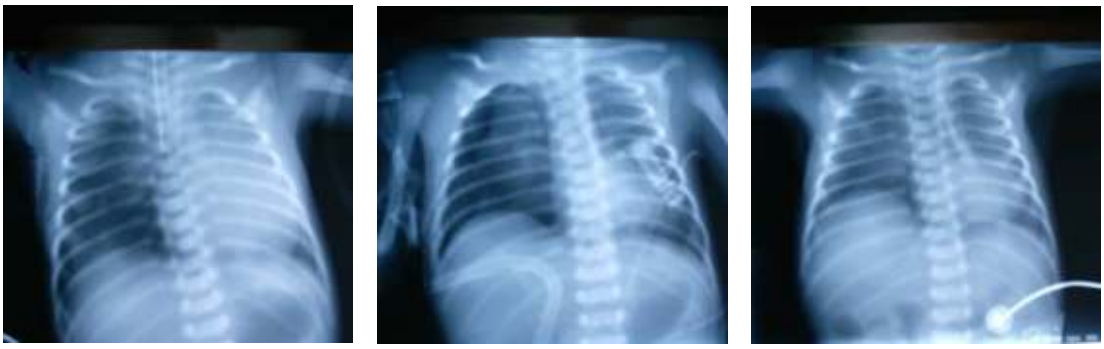

Fig. 2,3,4. Right pneumothorax and pneumomediastine

Starting the 2nd week of life, there was noticed a progressive increase of the cranial circumference, from 31,5 cm at birth to 34 cm ageing 2 weeks, 35 cm at 3 weeks of age and 40 cm at 8 weeks of age.

Transfontanellar ultrasound (baby girl aged 8 days): well designed cerebral tissue, right lateral ventricle 21 mm, left lateral ventricle 19 mm, with nonomogenous transsonic insights, anterior corns with hypereccogenous walls, moderately dilated 3rd ventricle with massa intermedia present. All of the above stand for evolutive bilateral hydrocephalus (moderate form). No signs of intraventricular hemorrhage.(fig.5)

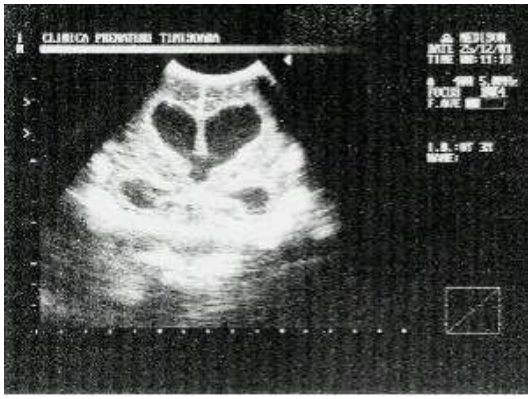

Fig. 5. Transfontanellar ultrasound – no sings of intraventricular hemorrhage.

Hydrocephalus is most frequently due to Sylvius's aqueduct stenosis, obstruction of the foramina of Luschka or Magendie. It is seldom related to an increase secretion of cerebral spinal fluid, to a decreased reabssorbtion in the subarachnoid space or to an arterial-venous malformation. Isolated hydrocephalus is more often caused by a congenital obstruction of Sylvius's aqueduct which makes the connection between the 3rd and the 4th ventricles. Hydrocephalus is a common feature of meningocele associated with Arnold Chiari malformation. When detected postnatal, the hydrocephalus might be following a bleeding process or a cerebral infectious episode.

First evaluation:

- Premature baby girl (gestational age 33 weeks)
- Caesarean section for bordered placenta praevia
- Respiratory distress syndrome-neonatal transient tachypnea
- Pneumothorax secondary to the therapy and to the right frenic nerve paralysis
- Evolutive hydrocephalus starting the 2nd week of life; first considered as a post bleeding process and then confirmed by the lumbar puncture (cerebral spinal fluid analysis) and the transfontanellar ultrasound

Question to be raised at this point: <u>early postnatal evolving hydrocephalus associated to the unilateral frenic nerve paralysis might conclude to a complex syndrome?</u> In order to answer to this question (baby's age being 3 weeks), it was asked for a magnetic resonance imagery (severe

ecstasies of the intracerebrum liquid spaces on bilateral overtentorium level with obvious compression on the cerebrum tissues; possible identification of an arterial-venous malformation of approximately 3/3/2 cm at the cortico / subcortical level, just anterior to the frontal corn of the right lateral ventricle; drainage vessels and bloody regional clots- using T_1 hypo signal, T_1 hyper signal)(fig.6)

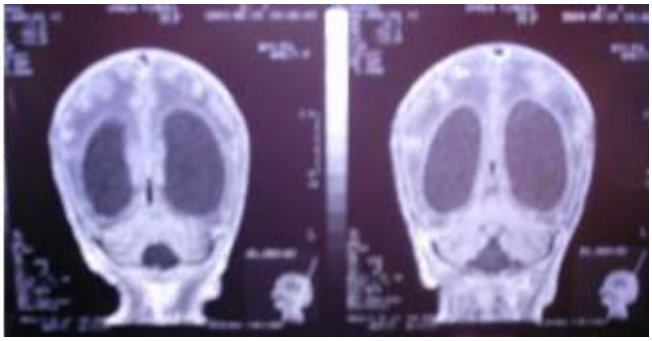

Fig. 6. Magnetic resonance aspect.

- Hypoplasic inferior cerebellum's vermix with moderate ecstasies of the 4th ventricle and it's large communication with cisterna magna; low insertion of the tentorium; intracerebral vessels- slightly ecstasies, sinuous shaped; hypophysis gland and orbits-normal (form of Dandy-Walker Syndrome)

Conclusions

1. severe triventricular hydrocephalus
2. right arterial-venous malformation with bloody regional clots (delayed sub acute status)
3. possible form of the Dandy-Walker Syndrome – partially agenesis of the anterior and inferior sector of the cerebellum's vermix
4. postnatal diagnosis via magnetic resonance imagery
5. standard incidences on the prenatal ultrasound scans of the cerebellum **DO NOT IDENTIFY** this possibility
6. an increasing need of adopting new incidences over the cerebellum while performing the prenatal ultrasound scan which might be borrowed from the magnetic resonance techniques (including transverse and oblique subcerebellar incidences)
7. **THE TRANSFONTANELLAR ULTRASOUND HAS IT'S OWN LIMITATIONS** related to the equipment itself, the techniques used, the examiner's abilities and of course the anatomical peculiarities identifiable in every baby at a certain age.

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