Nonimmune hydrops fetalis

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Abstract

BACKGROUND: Nonimmune hydrops fetalis (NHF) is an abnormal accumulation of fluid – especially serous – in visceral cavities and soft tissues. This condition may be caused by: cardiovascular diseases, chromosomal disorders, infections, lung, stomach, intestinal, kidneys, urinary tract and blood diseases, metabolic disorders and tumors. NHF may be diagnosed by an ultrasound scan.

THE AIM of the study was to present diagnostic and therapeutic difficulties as well as management with reference to NHF.

CASE STUDY: An abnormal accumulation of fluid in visceral cavities and subcutaneous tissue of two fetuses was diagnosed by an ultrasound scan. Despite a detailed and specific diagnostic procedure which included: infections, congenital malformations, chromosomal abnormalities etc. it was impossible to establish the cause of NHF. The symptomatic therapy was performed: periodic cordocentesis with an injection of human albumin solutions. In case of the first fetus therapeutic thoracocentesis was performed. The fetuses were delivered in 32nd and 31st week of pregnancy. Both neonates survived but even after the delivery it was impossible to establish the cause of NHF.

CONCLUSIONS: Multidirectional diagnostic approach is essential for the implementation of causal treatment of NHF. In case of idiopathic NHF the only management is symptomatic therapy, fetal monitoring and preterm delivery.

INTRODUCTION

Fetal hydrops is an abnormal accumulation of fluid – especially serous – in visceral cavities and soft tissues. In the past this condition was most often diagnosed in case of Rhesus (Rh) isoimmunization of the mother. Recently such cases have been occasional by reason of common prophylaxis consisting in anti-D immunoglobulin administration. However, the problem of nonimmune hydrops fetalis (NHF) caused by other reasons than Rh isoimmunization still remains. The cause of such condition may be as follows: cardiovascular diseases, chromosomal disorders, infections, lung, stomach, intestinal, kidneys, urinary tract and blood diseases, metabolic disorders and tumors. Nowadays, the high definition ultrasound machines make the diagnosis easy, however, a great diversity of possible causes of NHF still pose a diagnostic and therapeutic difficulty. Due to the NHF perinatologists are frequently impelled to induce a preterm labour in cases when continued gestation represents a higher risk for the fetus than a preterm delivery.

The aim of the study was to present, on the basis of two cases, diagnostic and therapeutic difficulties and management of NHF.
was performed in 32nd week of pregnancy (Velemín-
of ascending chorioamnionitis, a caesarian section
preterm premature rupture of membranes and the risk
as prophylaxis. Due to re-increase of pleural effusion,
Inflammation markers were negative
fluid index (AFI) according to Phelan stayed within
In 30th week of pregnancy preterm premature rupture
injected. The therapy was performed again in 26th,
The therapy was performed again in 24th
karyotype. Due to an accumulation of fluid in pleural
cavities of the fetus the diagnostic and therapeutic tho-
centesis were performed. The result of fluid culture
was negative, chylothorax was excluded, no DNA indi-
cating CMV, Toxoplasma or B19V infection was pres-
ent. After interim recovery re-accumulation of the fluid
in the pleural cavities of the fetus was observed. In the
22nd week of pregnancy it was decided to perform again
the thoracocentesis and cordocentesis. The fetal blood
cell count and biochemical parameters were normal
excluding decreased level of total serum protein (2.54 g/
dl) and albumin (2.03 g/dl). Therefore, human albumin
solution was injected to the umbilical vein resulting in
de edema decrease. During next two weeks slow accumu-
lation of fluid in subcutaneous tissue, peritoneum and
pleural cavities of the fetus was observed. Therefore
cordocentesis was performed again in 24th week of
pregnancy. Fetal blood cell count and biochemical parameters were similar to the
previous ones and human albumin solution was again
injected to the umbilical vein. The therapy was imple-
mented in 26th, 28th and 30th week of pregnancy due
to increasing edema and each time resulted in periodic
recovery. In 31st week of pregnancy cardiocotographic records and the umbilical artery and middle cerebral
tery Doppler indices revealed signs of fetal distress. A
premature newborn female of 2750g was delivered by a
cesarean section and obtained 6 points of Apgar score.
The neonate survived but the cause of NHF remained
unknown.

DISCUSSION
NHF occurs in 1:2500 to 1:3500 pregnancies (Faure et
al. 2004). Ultrasound markers of this condition are: skin
edema (fluid layer up to 5 mm wide, usually accumu-
lated in the area of head and neck), pleural effusion,
pericardial effusion (over 2 mm), ascites, placental
edema (placenta over 5 cm thick), abnormal amount of
amniotic fluid (usually polyhydroamnios, in some cases
oligohydramnios) (Merz, 2004).

The initial stage of NHF may differ. In our first case
the initial sign of NHF was pleural effusion whereas in
the second one skin edema appeared. Not all the symp-
toms of NHF listed above must appear, e.g. in both our cases placental edema and changes in the amount of amniotic fluid were not observed.

There are many pathophysiologic events resulting in abnormal accumulation of fluid in fetal compartments. Their recognition is of vital significance to differential diagnosis. The most frequent cause is the increased venous pressure as a result of failure of the cardiovascular system which is caused by different conditions (structural malformations, cardiac arrhythmia, serious anaemia, myocarditis) (Abrams et al. 2007; D’Amelio et al. 2006; Rose et al. 2005; Zeltser et al. 2003). The other cause is the decrease of the oncotic pressure resulting from lowered synthesis or an increased loss of proteins (congenital malformations, kidney and liver diseases) (Rodriguez et al. 2005; Ismail et al. 2001). The decreased level of proteins in the fetal blood may be the primary or secondary symptom of NHF, however it appears in most fetuses with NHF as a result of a chronic oxygen deficiency causing an increase in the permeability of vessels.

NHF etiology is heterogeneous and many factors may influence the final effect which is fetal hydrops. The number of factors to be considered makes the diagnosis complex and difficult. Nevertheless, the diagnostic process should never be abandoned as only the causal treatment may ensure long-lasting therapeutic effects. Despite many efforts, in 40% of cases it is impossible to identify the cause of NHF, as it was indicated in the cases described above (Merz, 2004). In such case the management consists in symptomatic therapy, fetal monitoring and a preterm delivery.

The mortality of neonates with NHF is high and varies from 60 to 81% (Abrams et al. 2007; Isaacs, 2008). In the cases described above our management resulted in the survival of the neonates.

Conclusions

Multidirectional diagnostic approach is essential for the implementation of causal treatment of NHF.

In case of idiopathic NHF the only management is symptomatic therapy, fetal monitoring and preterm delivery.

REFERENCES