Case report

PRENATAL INVASIVE DECOMPRESSION THERAPY OF AN ARACHNOID CYST IN A FETUS WITH NORMAL HEART ANATOMY AND NO FUNCTIONAL ANOMALIES - A CASE REPORT

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Abstract

We present a case of a female fetus with large posterior fossa cyst. After detailed diagnosis in referral center revealing normal heart anatomy and no functional abnormalities in cardiovascular system, a trial decompression of the fluid reservoir was suggested to the parents and successfully performed at the 30th week of pregnancy. Neurosurgical treatment was continued in the neonatal period. At the age of 7 months, the child presented normal physical development, and the cranial image of the CNS showed good recovery of the brain.

Key words: *arachnoid cyst, posterior fossa cyst, echocardiography, invasive decompression therapy of an arachnoid cyst*

INTRODUCTION

Existing literature on several occasions points to the role of a fetal cardiac examination in the qualification for invasive procedures, these include anomalies of the chest and ovarian cysts[1,2,3]. Nevertheless, so far no reports have been published on qualification for prenatal arachnoid cyst decompression.

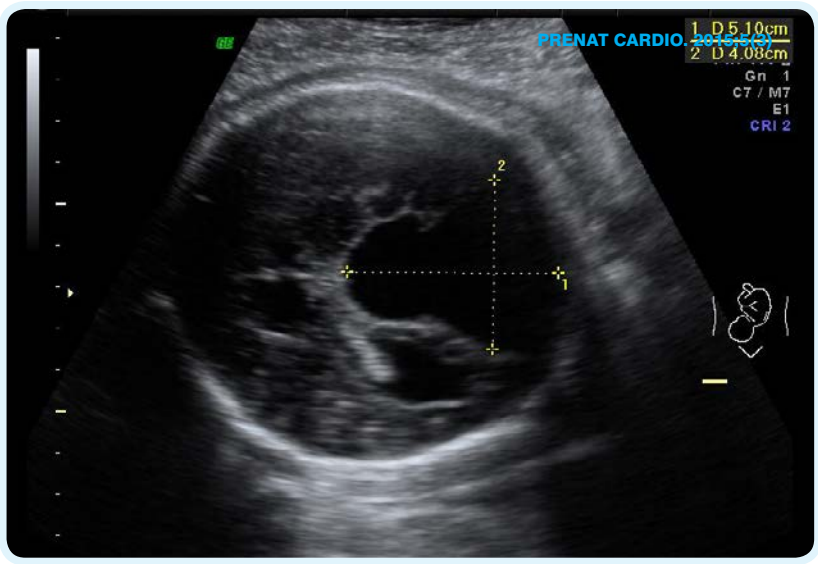

Figure 1. A cyst of the posterior fossa at 35.3 week of gestation: 5.1 x 4 cm.

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A CASE REPORT

A 23-year-old multigravida, second pregnancy, second labour (first pregnancy in 2009, caesarean section, healthy male newborn), reported to our centre at the 35th week of gestation because of the detection of a fluid reservoir in the posterior fossa of the skull. Initially, there was a suspicion of Dandy-Walker syndrome, but ultimately prenatal diagnosis based on targeted ultrasound allowing us to confirm the posterior fossa cyst measuring 5x4 cm, with secondary widening of the posterior corni of the lateral ventricles.

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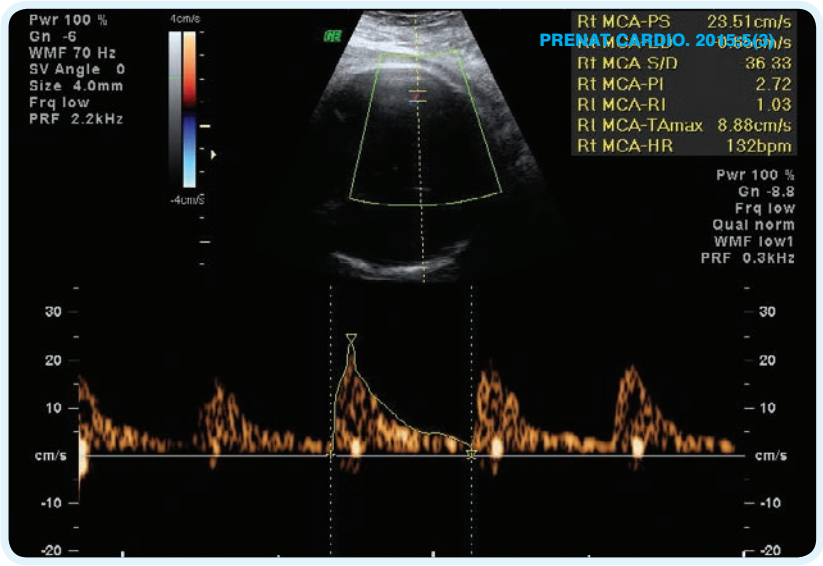

Figure 2. The normal pulsatility index of middle cerebral artery (PI 2,72)

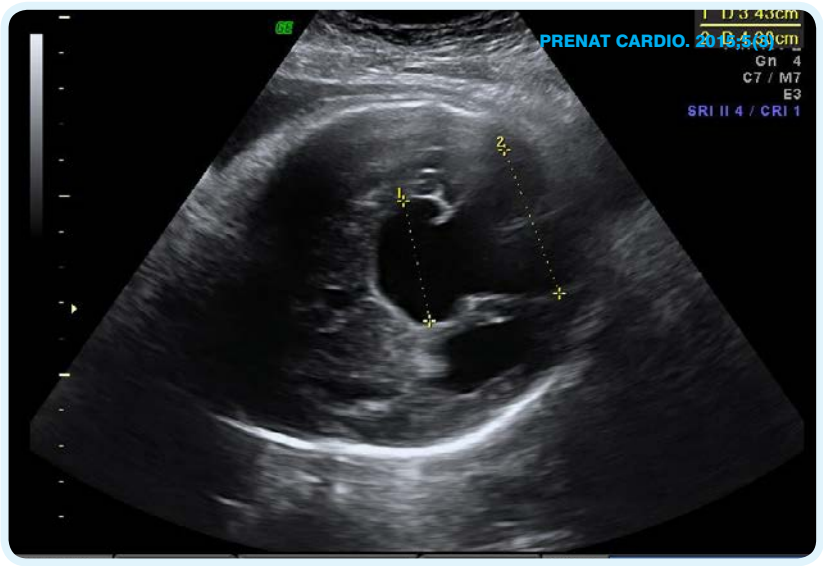

Figure 3. A cyst of the posterior fossa at 38.2 wk of gestation 3,4 x 4,3 cm

Fetal echocardiography confirmed normal heart anatomy and excluded functional changes in the cardiovascular system, which allowed for qualification to attempt intrauterine cyst decompression. 40 ml of colourless fluid was obtained during the procedure. Four weeks after the intervention the head size of the fetus was evaluated as normal despite the persistence of the cyst; however, its dimensions were diminished (3 x 4cm) and echocardiography reaffirmed the wellbeing of the fetus.

At 38th week, elective caesarean section was performed and female newborn was delivered with birth weight 3300g and Apgar score of 9 points.

MRI and CT-angiography confirmed the presence of a posterior fossa cyst measuring 70 x 61 x 49mm. Dandy-Walker syndrome was excluded and the newborn was qualified for Rickham reservoir implantation on the 9th day of life. The Rickham reservoir was punctured every day, firstly 10 ml of the cerebrospinal fluid was released, and then it increased to 20 ml/day. A cranial ultrasound still presented a cyst with dimensions of 42 x 46 x 40 mm with a slight enlargement of the lateral ventricles. On the 43rd day of life, in our institute, the Rickham reservoir was surgically removed and ventriculo-peritoneal Orbis-Sigma valve was implanted. The child was discharged from hospital in good general condition on the 55th day of postnatal life.

Control ultrasound + ECHO was performed in the infant at seven months. The structures of the CNS were described as normal, without evidence of a significant expansion of the ventricular system.

DISCUSSION

An arachnoid cyst is a rare congenital central nervous system defect with an incidence of about 1-2%, probably arising after the period of embryogenesis [4]. An arachnoid cyst is a fluid space between the separated walls of the arachnoid, which does not communicate with the subarachnoid space [4,5].

Arachnoid cysts can occur as primary or secondary. Primary is a consequence of abnormal embryogenesis of the arachnoid, and secondary (or acquired) results from bleeding, trauma or infection [4]. Secondary arachnoid cysts usually communicate with the subarachnoid space and by definition are not "real" arachnoid cysts [4].

Arachnoid cysts can develop anywhere along the arachnoid membrane. They usually occur as single-chamber changes with regular or irregular outlines, located in the midline or asymmetrically [4]. Their characteristic feature is usually the non-regular outline [4]. The cyst size ranges from a few to about 50 mm, which makes the cysts of a few millimetres often identified by chance in the postnatal period, and only in CNS imaging (CT or MRI), in children or adults [5].

The size of the cyst may at some point be constant or increase gradually with the developing fetus, causing secondary ventriculomegaly [6], as in the case presented in this study. Arachnoid cysts can occur as isolated defects or constitute part of a syndrome [7,8].

Most arachnoid cysts are asymptomatic by nature, but they can also be the cause of hydrocephalus and its complications [4,6]. Other consequences of cysts include: bleeding into the cyst, bleeding into the subdural chamber, and even rupture of the cyst [5].

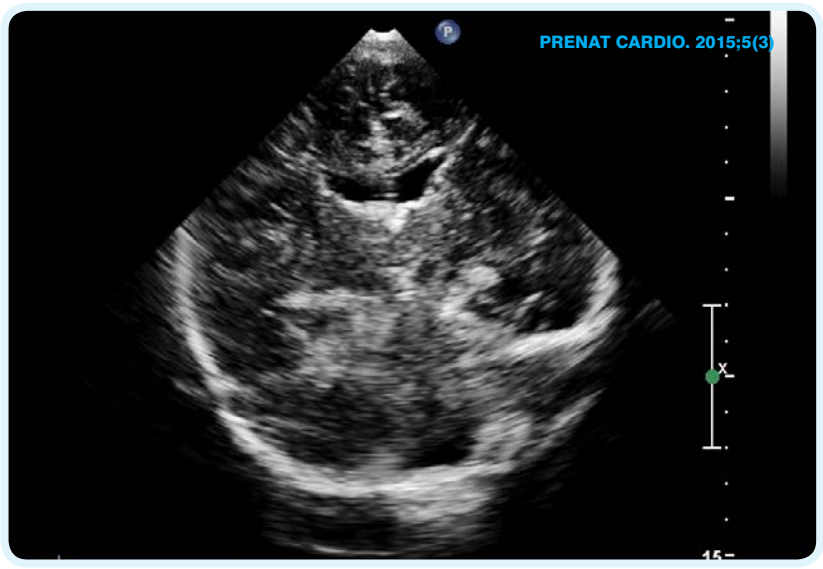

Figure 4. The cranial ultrasound at 7 months after prenatal and postnatal decompression of the cyst.

Treatment of arachnoid cysts is varied and controversial, it is usually dependent on symptomatology [9]. So far, except for individual descriptions of cases, no therapy in utero has been reported for arachnoid cysts; similarly, nobody emphasized the role of echocardiography in the qualification for decompression of cysts in utero.

Therapeutic strategies in the event of arachnoid cysts in children are well described and include waiting and observation, installation of a shunt and endoscopic or open fenestration of the cerebrospinal fluid from a cyst to the reservoirs of the base of the brain [9]. One of the ways of treatment is needle aspiration of the cyst [5]. However, if the cause of the cyst was the lack of drainage of the cerebrospinal fluid, a cyst can easily renew, which will result in the need for further aspirations. Another treatment option is craniotomy with excision of the cyst wall and fenestration of the cerebrospinal fluid to the reservoirs of the base of the brain [10]. An endoscopic method is less invasive, and involves the creation of the connection between arachnoid cysts and the reservoirs of the base of the brain or the ventricular system. The advantage of endoscopic methods include lower incidence of complications and negligible mortality [9]. Microsurgical method of the installation of a ventricular-amniotic shunt is estimated to be as safe and effective as the endoscopic method, however, it is more expensive and more complicated procedure. Also, the minimally invasive stereotactic method does not always allow for the creation of large and efficient connections [9].

In the literature regarding invasive procedures in prenatal life, the role of prenatal cardiac evaluation before surgery as an important factor, is limited. Based on our previous experience and data from literature we underline again the value of echocardiography in the qualification and conduction of intrauterine therapy in the cases of extracardiac anomalies, for instance hydrothorax, CALM, DH, ovarian cyst [1,3].

In our centre we have adopted a rule of disqualification from in utero treatments in non-cardiac defects when fetal heart defects coexisted [1,2].

In the case of normal heart anatomy with no functional changes, fetuses would well tolerate both anaesthesia of the mother and surgery itself, which has been confirmed by this study.

The Polish Mother's Memorial Hospital Research Institute recommends serial fetal in utero echocardiography before and after surgery during the entire diagnostic-therapeutic process[1,2].

CONCLUSIONS

Prenatal invasive therapy for large arachnoid cyst in fetus with normal heart anatomy and no haemodynamic abnormalities was the first step in a multi-stage treatment, ending with the successful development of the infants CNS structures.

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Mikołajczyk-Wieczorek W: neurosurgeon data collection, work with manuscript

Słodki M: concept of the research, final version