



A case of non-immune hydrops fetalis with cystic hygroma complicated by intrauterine fetal death

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Abstract

Hydrops fetalis refers to the excessive accumulation of fluid in the extracellular compartment of the fetus. Cystic hygroma is a congenital malformation of the cervical lymphatic system responsible for an accumulation of lymphatic fluid commonly in the retrocervical region. Cystic hygromas are most often associated with chromosomal abnormalities. When a cystic hygroma is diagnosed in utero, the fetal survival rate is only 2-6%. When hydrops fetalis is present with cystic hygroma, the mortality rate is close to 100%. We present here the case of a 23-year-old primigravida in whom an ultrasound performed at 17 weeks of amenorrhea revealed a posterior cervical cystic hygroma associated with hydrops fetalis, the evolution of which was marked by intrauterine fetal death two weeks later.

Keywords: *Hydrops fetalis, Cystic hygroma, Chromosomal abnormalities, Prenatal diagnosis.*

Introduction

Hydrops fetalis was first described in 1982 by Ballantyne. It is a diffuse subcutaneous oedema, associated at least with an effusion at the level of a serosa: pericardium, pleura, peritoneum^[1]. It can also be defined by the presence of an effusion in two different serosa. A polyhydramnios is often associated, but it is fickle and therefore not part of the definition. Thus, depending on the etiology, hydrops is associated with polyhydramnios,

oligohydramnios or a normal amount of amniotic fluid.

Hydrops fetalis results from an imbalance in water homeostasis, with accumulation of fluid in the extracellular compartment. This imbalance can result from two main categories of pathologies. This is how we distinguish immune hydrops fetalis, by fetomaternal blood incompatibility (most often in the rhesus D system), from non-immune hydrops fetalis. There are more than 100

recognized etiologies of non-immune hydrops fetalis^[2].

Cystic hygroma is a congenital malformation of the jugular lymphatic system which presents as a compartmentalized cystic sequestration of lymphatic fluid, most often at the retro-cervical level.

If a connection between the lymphatic system and the venous system does not occur at this stage, the evolution will be marked by hydrops fetalis, which will lead to early intrauterine fetal death.

Case Report

We report a case of a 23-year-old patient, A RhD positive blood (A+), primigravid nulliparous, with no history of diabetes or consanguineous marriage.

The first contact in our department dates back to 17 weeks of amenorrhea when an obstetric ultrasound objectified a progressive intrauterine monofetal pregnancy with cystic hygroma (Figure 1 and 2) and hydrops fetalis (Figure 3), with no other detectable fetal malformation.

The search for irregular agglutinins was negative, concluding that hydrops fetalis was non-immune.

A control ultrasound carried out 2 weeks later, at 19 weeks of amenorrhea, found an intrauterine fetal death. A wait of 1 month was proposed to the patient in view of a spontaneous expulsion of the product of conception, without success. At 23 weeks of theoretical gestational age, induction of labor by vaginal misoprostol at a dose of 200 µg every 6 hours, resulted after the 3rd pose in the expulsion of a stillborn in one piece, female, weighing 705 grams. The macroscopic examination found a cleft lip and cleft palate, a large cystic hygroma and a severe generalized edema (Figure 4). A fetal karyotype from a piece of unfixed umbilical cord would have been highly desirable in the search for chromosomal anomalies, but our country does not currently have structures with the technologies necessary to carry out this examination. A fetal autopsy was proposed but the couple refused to perform it.



Figure 1: Cystic hygroma, ultrasonographic transverse view.



Figure 2: Cystic hygroma, ultrasonographic sagittal view.



Figure 3 (A)



Figure 3: Ultrasonographic showing ascites (A) and hydrothorax (B)



Figure 4 : Aborted female fetus presenting large cystic hygroma, severe generalized edema, with cleft lip and palate

Discussion

➤ Physiopathology

- ❖ Cystic hygromas result from abnormal development of the posterior cervical lymphatic network, accompanied by abnormal budding of the lymphatic tissue

with sequestration of lymphatic fluid and absence of a venous outlet^[3].

➤ Location

- ❖ The most common location of cystic hygroma is the posterior cervical region, which accounts for 80% of locations^[4]. Other locations include the cervico-occipital region as well as the laterocervical region.

➤ Associations

- ❖ Cystic hygromas are associated with chromosomal abnormalities in 60 to 75% of cases depending on the series. The chromosomal abnormality most frequently associated with cystic hygroma is monosomy X or Turner syndrome (45,X)^[5]. Note also Trisomy 21, 18 and 13, as well as Klinefelter syndrome (47, XXY)^[6]. Furthermore, intrauterine exposure to alcohol has also been associated with the development of cystic hygroma.

➤ Positive diagnosis

- ❖ The diagnosis of cystic hygroma is made by ultrasound. It is characterized by an anechoic posterior cervical cystic mass, comprising at least two cysts separated by a septum. Its size is variable^[7].

➤ Differential diagnosis

- ❖ There are two main differential diagnoses. Cystic hygroma must be distinguished from occipital meningoencephalocele (presence of an occipital bone defect and intracranial communication), and from cystic lymphangioma of the neck (which is more anterior and lateralized)^[8].

➤ Prognosis

- ❖ It depends on the presence or absence of chromosomal abnormalities. Cystic hygromas without chromosomal abnormalities tend to regress and give way to a thick nape in the second trimester of pregnancy, then to pterygium colli in the postnatal period^[9].
- ❖ On the other hand, the prognosis is poor in case of associated chromosomal

abnormalities. Intrauterine fetal death usually occurs shortly after diagnosis. It is due to chronic fetal hypoxemia secondary to compression of the thoracic structures by generalized edema. When hydrops fetalis is associated with cystic hygroma, the mortality rate is close to 100%^[10].

Conclusion

This case report highlights the association between cystic hygroma and hydrops fetalis. The diagnosis of cystic hygroma is made by ultrasound. Cystic hygroma can be complicated by pleural and pericardial effusions, ascites or even hydrops fetalis, often progressing to spontaneous termination of pregnancy at the beginning of the second trimester. Chromosomal abnormalities, mainly aneuploidies, are frequently associated with cystic hygroma justifying the realization of a fetal karyotype for etiological purposes.

Declaration of parental consent: The authors certify that they have obtained consent from the parents.

Conflicts of Interest: The authors declare that they have no conflict of interest.

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