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CASE REPORT

Diagnosis of Non-Immune Hydrops Foetalis and Cystic Hygroma in a 20-Week Foetus: A Case Report

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Summary

Hydrops foetalis (HF) is the excessive fluid accumulation in at least two foetal body cavities, including pleural, pericardial, and peritoneal cavities, with associated soft tissue oedema. Most cases of hydrops foetalis were due to severe erythroblastosis foetalis, secondary to Rhesus isoimmunization. However, other variants include non-immune hydrops foetalis (NIHF). Foetal cystic hygroma, which occurs due to lymphatic obstruction, is commonly associated with immune hydrops foetalis (IHF). A combination of the conditions in a foetus tends to portend an abysmal prognosis because it can lead to abortion, intrauterine foetal death (IUFD) and early neonatal death (ENND). We report a case of a 22-year-old woman with a prenatal ultrasound diagnosis of a combination of NIHF and foetal cystic hygroma at 20 weeks of gestation. Therefore, there is a need for early diagnosis of the anomalies so that early termination of pregnancy can be performed if the patient desires.

Key words: Antenatal Care, Cystic hygroma, Foetus, Non-immune Hydrops foetalis, Ultrasound scan.

Introduction

Hydrops foetalis (HF), known as 'oedema of the foetus' in Latin, is the presence of extracellular fluid in at least two foetal compartments. The commonly affected sites and compartments are the skin and subcutaneous tissue, pleural cavity, pericardial cavity, and peritoneal cavity. [1]

There are two broad groups of HF: immune HF (IHF) and non-immune HF (NIHF). The non-immune types account for the greater percentage of foetal hydrops. [2] The aetiology of IHF is foetal erythroblastosis from Rhesus incompatibility between the mother and the foetus. In 50% of cases, NIHF is idiopathic but aetiological factors such as foetal cardiac anomalies, twin-to-twin transfusion syndrome, Turner syndrome, Trisomies (13, 18 and 21), renal diseases, placental tumours, and parvovirus infections have also been reported. [1, 2]

Foetal cystic hygroma occurs because of early lymphatic obstruction. Its appearances range from increased foetal nuchal translucency to multiple thin-walled cystic masses around the foetal head and neck. [3] Cystic hygroma with NIHF has been associated with a high incidence of aneuploidy and increased mortality. [3, 4]

This report describes a 22-year-old woman who had a prenatal diagnosis of NIHF and cystic hygroma at 20 weeks of gestation. The pregnancy was terminated after counselling, and the anomalies were grossly seen and confirmed.

Case Description

A 22-year-old G_2P^{1+0} (1 alive) woman, unsure of her last menstrual period, was referred from a primary healthcare centre to the Ultrasound unit of the University College Hospital, Ibadan, Nigeria, for a detailed foetal anomaly ultrasound scan. An earlier ultrasound scan at 20 weeks at a private facility showed foetal ascites. The pregnancy was conceived spontaneously three months after removing a subdermal contraceptive implant, which the mother had for three years. Following the removal, her menstrual flow was irregular, which made her unsure of her last menstrual period. She had carried a pregnancy to term and had a spontaneous vertex delivery of a healthy baby three years before the index pregnancy.

A detailed foetal anomaly ultrasound scan at our facility showed a live singleton intrauterine foetus at an approximate gestational age of 19 weeks and five days. There was generalized oedema evidenced by thickened skin and subcutaneous tissues over the foetal scalp, the abdomen, and the limbs. There were also foetal ascites, pleural effusion, and pericardial effusion. (Figures 1 and 2)

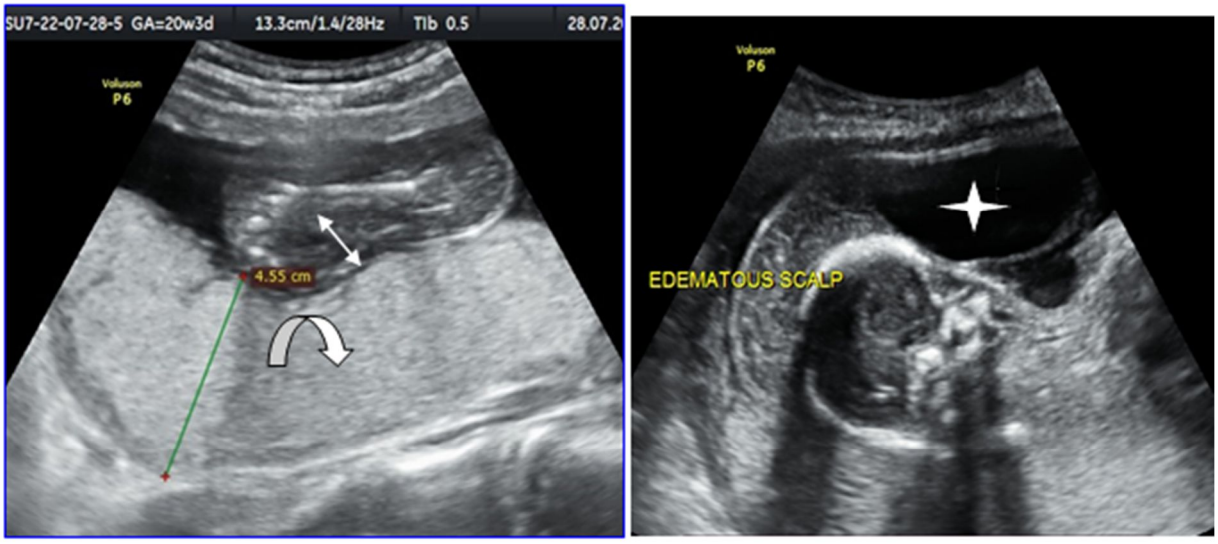


Figure 1: 2D-Ultrasound images of the foetus showing posterior placenta (curved arrow) with a maximal thickness of 4.5cm. It also shows subcutaneous oedema on the foot (white double arrow) and the scalp. A cystic mass (star) is seen on the side of the head consistent with cystic hygroma.

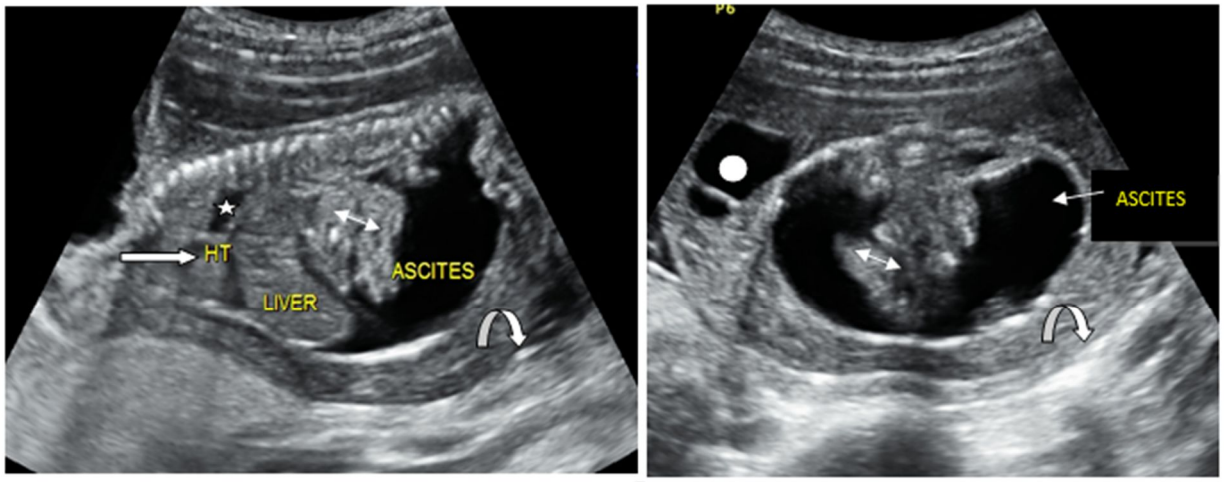
A huge multiloculated cystic mass was also seen around the foetal head and neck, suggestive of cystic hygroma. The largest of the locules measured 6.5cm in its widest diameter (Figure 3), and there was no vascularity on colour Doppler interrogation. However, the heart showed no obvious abnormality.

The placenta was located posteriorly with a thickness greater than 4cm, consistent with placentomegaly in relation to the gestational age. Still, the amniotic fluid volume was within normal limits. An ultrasound impression of Hydrops foetalis with an associated cystic hygroma in a live 19 weeks, five days foetus was made. The patient was counselled on the ultrasound findings and possible diagnosis and prognosis, and she opted for medical termination of pregnancy. She was subsequently admitted into the gynaecological ward. Her packed cell volume and urinalysis were within normal limits. Her blood group was O, 'Rh' D^+, while her husband was A, 'Rh' D^+. The retroviral screening was not reactive at the time of screening, and the hepatitis B surface antigen was also negative.

Medical termination was instituted with 4-hourly insertion of 400mcg of misoprostol into the posterior fornix of her vagina. She expelled the products of conception 10 hours after two doses of misoprostol. On examination, the product of conception was a malformed baby with generalized oedema and bilateral cystic neck masses (Figure 4). She was then counselled on preconception care before her subsequent pregnancy and the need for prenatal care in her next pregnancy. She was discharged home on oral folic acid for three months.

Patient's perspective: The patient was glad she terminated the pregnancy after seeing the product of conception. She promised an early antenatal booking in subsequent pregnancies in a tertiary hospital where an anomaly ultrasound scan would be done in all pregnancies.

Informed consent: The authors certify that they obtained appropriate consent from the patient to use the images and other clinical information in this journal article. The patient understood that her identity would not be revealed.



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Figures 2a and b: Ultrasound images of the foetus (a) sagittal view (b) axial view – showing pleural effusion (star), pericardial effusion (thick arrow) and ascites, with the bowels (white double arrow) floating within it. Skin oedema (curved arrow) and cystic hygroma (circle) are also noticed.

Discussion

A combination of hydrops foetalis and cystic hygroma on prenatal ultrasound scans, especially in the second trimester, has been associated with grave prognoses such as abortion, intrauterine foetal death and early neonatal death. [5, 6]

An ultrasound diagnosis of hydrops foetalis is made when there is excessive accumulation of fluid in at least two different foetal compartments with or without generalized subcutaneous oedema, and the excess fluid may accumulate within the pleural, peritoneal, or pericardiac cavities. [2] Other sonographic features in HF include polyhydramnios, increased placental thickness, and enlarged liver and spleen. [1,2] In the index case, generalized oedema, excess fluid in all the body cavities, and increased placenta thickness were noted.

Hydrops foetalis can be broadly categorized into immune and non-immune HF. The diagnosis of immune HF is made when there is Rhesus incompatibility between the mother

and the foetus, while a non-Immune aetiology is suspected when there is no incompatibility. [1,2] Non-immune hydrops foetalis is a rare condition with a wide incidence, ranging between 1 in 1500 and 1 in 4000 worldwide. The variation in the prevalence may be due to differences in definition, population, and the thoroughness of evaluation. [3] In the index case, there was no obvious evidence of Rhesus incompatibility because the mother was Rhesus D positive, which made NIHF a likely diagnosis.

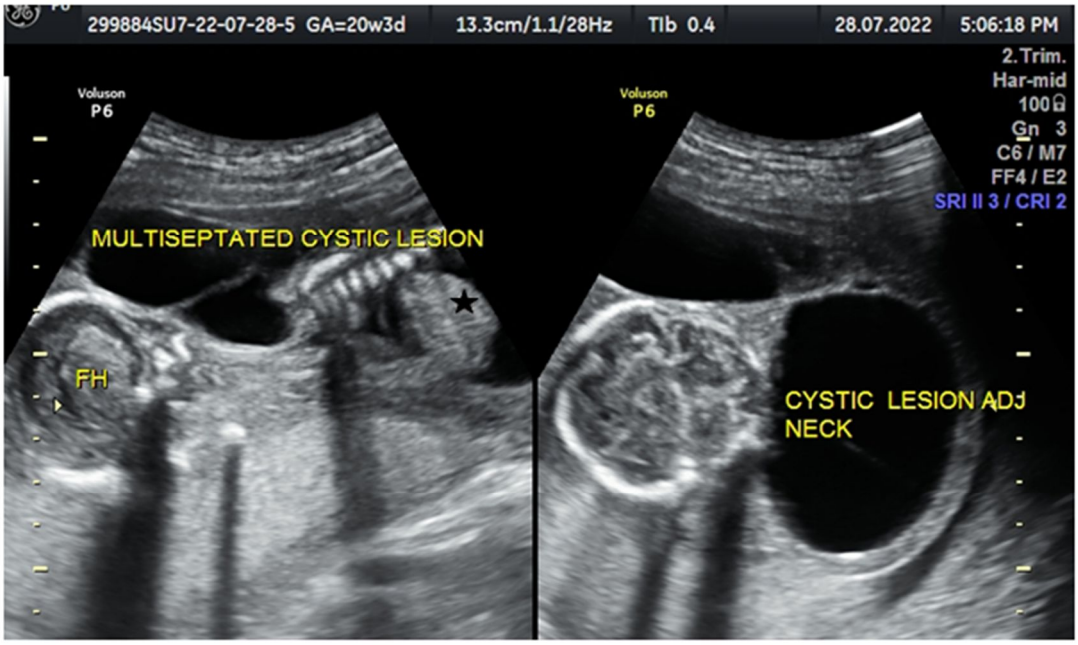

Figure 3: 2D-Ultrasound of the foetus in (a) longitudinal and (b) transverse views showing a huge multiloculated cystic mass around the foetal head and neck consistent with cystic hygroma.

Conversely, cystic hygroma is a congenital obstruction of the foetal lymphatic drainage. The ultrasonographic diagnosis is made where cystic masses are seen at the cervicofacial and axillary regions of the foetus. The sonographic appearances of these cystic masses may be septated or non-septated.[4] When the cystic mass is small, it usually regresses spontaneously. However, a large cystic hygroma rarely regresses and is generally associated with hydrops foetalis. In isolated cases of cystic hygroma diagnosed in-utero, the foetal survival rate is 2–6%, while the mortality rate is almost 100% when combined with HF. [6, 7] In the index case, the cystic masses were large and septated, seen around the foetal head and neck with associated HF.

The high mortality rate observed in a combination of cystic hygroma and NIHF has been linked to the increased incidence of foetal chromosomal anomalies. The most frequently associated chromosomal anomaly in the first trimester is Trisomy 21, followed by Turner syndrome (45X) and Trisomy 18. However, in the second trimester, Turner syndrome (45X) is the commonest until proven otherwise. Turner syndrome has a variable presentation, but NIHF with cystic hygroma in-utero is the most severe form, with high mortality. [7-10] In the index case, the diagnosis was made in the second trimester. Therefore, there is a strong likelihood that it may be due to Turner syndrome.

Genetic counselling with foetal karyotyping should be offered to affected parents to confirm

the diagnosis of foetal chromosomal anomalies. However, many studies have reported poor prognoses, even in patients with normal karyotypes. [8-10] In this index case, karyotyping was not offered because the mother might not be able to afford it because of her low socioeconomic status, which made her register at a primary health centre for antenatal care.

Due to the high mortality rate associated with HF and cystic hygroma, pregnant women are usually counselled after making the diagnosis and allowed to terminate the pregnancy if they so desire. [8, 9] The pregnant woman in the index case was adequately counselled and opted to terminate the pregnancy.

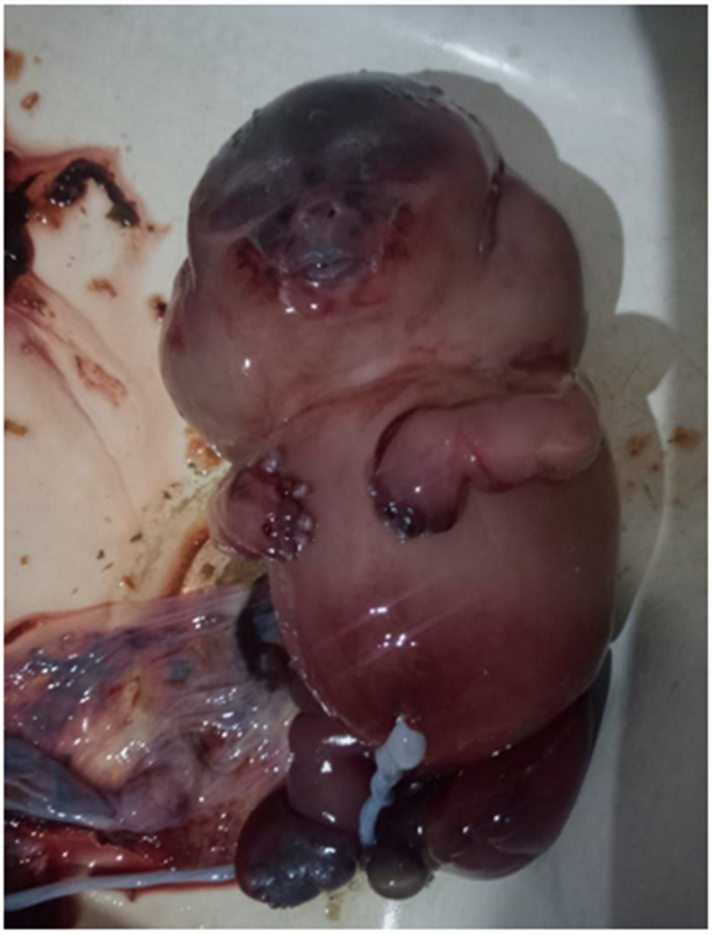

Figure 4: Image of the post-abortal foetus showing the generalized body swelling (anasarca) and the large neck mass (cystic hygroma).

Conclusion

The co-existence of hydrops foetalis and cystic hygroma in the second trimester has been associated with abortion, IUFD and ENND. Therefore, there is a need for early diagnosis of the anomalies so that early termination of pregnancy can be performed if the patient desires.

Authors' Contribution: AJA and FOE conceived the study. FOE drafted the manuscript. All three authors did a literature review, revised the draft for sound intellectual content and approved the final version of the manuscript.
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