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Case Report

Anencephaly in a triplet pregnancy: Unprecedented spontaneous reabsorption in-utero and subsequent normal delivery via c-section: A rare case report ☆

Amrit Bhusal, MBBS[a,*], *Tek Nath Yogi, MBBS*[b], *Jwala Budthapa, MD*[a], *Shailendra Katwal, MD*[c], *Asim Mahat, MD*[d]

[a] *Department of Radio-diagnostics and Imaging; BP Koirala Institute of Health Sciences (BPKIHS), Dharan, Sunsari, Nepal*
[b] *BP Koirala Institute of Health Sciences (BPKIHS), Dharan, Sunsari, Nepal*
[c] *Dadeldhura Subregional Hospital; Dadeldhura, Nepal*
[d] *Department of Radiodiagnosis and Imaging; Nepalese Army Institute of Health Sciences, Kathmandu, Nepal*

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ABSTRACT

Multiple pregnancies are infrequently encountered, with the incidence of spontaneous triplet pregnancies estimated at approximately 1 in 7000 pregnancies. Triplet gestations are recognized for their propensity to bring about a spectrum of pregnancy related complexities, encompassing fetal structural abnormalities, neurological anomalies, disturbances in amniotic fluid levels, preterm labor, and suboptimal neonatal outcomes. Anencephaly is a serious congenital defect where the brain and skull do not fully develop, often leading to a poor prognosis. It's a preventable neural tube defect (NTD) with timely intake of folic acid, is caused by the incomplete closure of the neural tube during fetal development, resulting in the absence of the cerebrum (responsible for thinking and coordination) and the front part of the brain (forebrain) in affected infants. While anencephaly in a triplet is scarcely reported, spontaneous reabsorption of an anencephalic fetus in utero is a rare and unexpected event, with no documented cases in triplet pregnancies until now. We report a case of anencephaly in a triplet pregnancy where the mother presented late during her third trimester, the reabsorption of the anencephalic fetus in utero is an unprecedented event, highlighting the unique nature of this triplet pregnancy.

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* Corresponding author.
E-mail address: Amritbhusal51@gmail.com (A. Bhusal).

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Introduction

Multiple pregnancies are infrequently encountered, with the incidence of spontaneous triplet pregnancies estimated at approximately 1 in 7000 pregnancies. Triplet gestations are recognized for their propensity to bring about a spectrum of pregnancy-related complexities, encompassing fetal structural abnormalities, neurological anomalies, disturbances in amniotic fluid levels, preterm labor, and suboptimal neonatal outcomes [1].

Furthermore, it is imperative to acknowledge that triplet pregnancies exhibit an elevated predisposition to the occurrence of congenital anomalies, such as anencephaly. Anencephaly is a serious congenital defect where the brain and skull do not fully develop, often leading to a poor prognosis. It's a preventable neural tube defect (NTD) with timely intake of folic acid, is caused by the incomplete closure of the neural tube during fetal development, resulting in the absence of the cerebrum (responsible for thinking and coordination) and the front part of the brain (forebrain) in affected infants [1,2].

The diagnosis of anencephaly can be made through routine transabdominal ultrasound examination, as early as 11 weeks of gestation after which diagnostic challenges may occur due to complications and differences in radiological features in the second and third trimesters. While anencephaly in a triplet is scarcely reported, spontaneous reabsorption of an anencephalic fetus in utero is a rare and unexpected event, with no documented cases in triplet pregnancies until now. It poses a significant risk to the lives of both the mother and the fetus [2–4]. In this rare case of anencephaly in a triplet pregnancy where the mother presented late during her second trimester, the reabsorption of the anencephalic fetus in utero is an unprecedented event, highlighting the unique nature of this triplet pregnancy.

Case presentation

A 34-year-old female, initially presented at 25 weeks of gestation with a confirmed triplet pregnancy in sonographic examination. She was otherwise healthy, with an unremarkable medical history. She did not give any history of prior medical checkups and any medication intake like folic acid. She gave history of spontaneous conception. At the time of her first antenatal visit, the patient underwent routine transabdominal ultrasound examination, which revealed the presence of triplets in the uterine cavity. Subsequent assessments of the fetuses identified a gross congenital anomaly—specifically, anencephaly—in 1 of the 3 fetuses. Transabdominal ultrasound image showed triplet pregnancy with normal flow in color Doppler study (as shown in the Fig. 1B), also showed triplet pregnancy with 3 separate placentae with absent cranial bone with bulging orbit giving "Frog eye appearance" (as shown in the Fig. 1A). This condition was characterized by the underdevelopment of the brain and skull, typically carries a poor prognosis. Her laboratory findings were unremarkable except for the rise in the level of alpha-fetoprotein.

After the initial diagnosis of the anencephalic fetus, the patient's follow-up appointments were scheduled to closely monitor the progress of the remaining two healthy fetuses. Unfortunately, the patient was lost to follow-up for an extended period of time. The patient presented in the third trimester, during the third trimester (32 weeks of gestation) of her pregnancy, seeking medical attention due to symptoms of premature labor. What emerged during subsequent evaluation was astonishing: the anencephalic fetus had undergone spontaneous reabsorption in utero (Figs. 2 and 3), a rare and unexpected event. This marked the first documented case of such an occurrence in a triplet pregnancy. The patient received counseling about her triplet pregnancy, where the chances of carrying all 3 fetuses with an anencephalous fetus to term were low. She was recommended for fetal reduction at the feto-maternal unit, but the patient chose to proceed with the pregnancy without reducing the number of fetuses.

Given the advanced gestational age and the potential risks associated with a triplet pregnancy, a decision was made to perform a cesarean section (LSCS) to ensure the safety of the remaining two viable fetuses. The procedure was executed without complications, leading to the birth of two healthy neonates.

Discussions

Incidence and challenges of triplet pregnancies

Multiple pregnancies have been attributed to 2 main factors, having conception at an older maternal age where multifetal gestations are more likely to occur naturally, and the increased use of assisted reproductive technology. Triplet pregnancies, while rare, pose significant maternal and fetal complications. With increased use of assisted reproductive technologies, their incidence may rise, necessitating improved obstetric care [4]. In a study by Esike et al. [5], 22 triplet pregnancies over a decade showed a 0.1% incidence. Most women were 31-35 years old, half being grand multiparas. About half delivered at 35-37 weeks, with 31.8% at term and 18.2% between 28 and 34 weeks. Key complications included preterm labor (22.7%), preeclampsia, prolonged rupture of membranes, polyhydramnios, and anemia. Perinatal mortality was 91 per 1000 deliveries, with 81.8% delivered vaginally and 18.2% via C-section. These findings underscore the complexities and risks associated with triplet pregnancies.

Anencephaly in triplet pregnancy

Anencephaly,a serious neural tube defect has been reported to be frequently associated with twin pregnancy while its occurrence in triplet pregnancies is rarely reported in literatures which has been addressed in the present case [4,6]. Multiple pregnancies can pose a greater risk of congenital anomalies in the growing fetus among which anencephaly is the common and serious neural tube defect. It occurs when unprotected brain tissue is exposed to the amniotic fluid, which can cause gradual brain tissue degradation due to mechanical damage, including friction with the uterine wall, placenta, and fetal

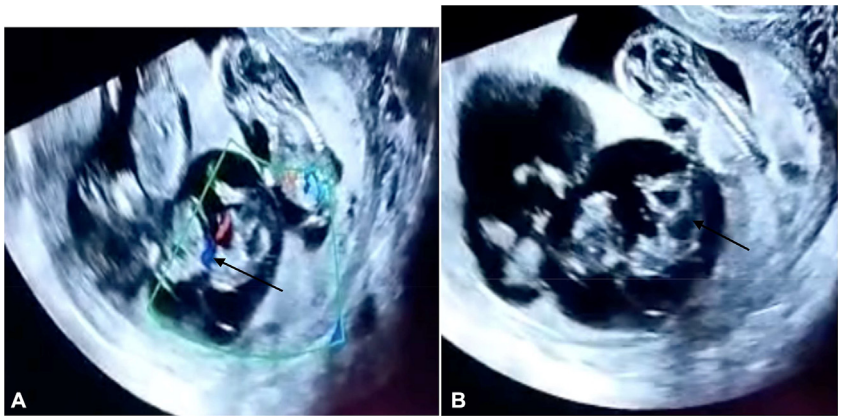

Fig. 1 – (A) Transabdominal ultrasound image showing triplet pregnancy with three separate placentae with absent cranial bone with bulging orbit giving "Frog eye appearance" (black arrow). (B) Transabdominal ultrasound image showing triplet pregnancy with normal flow in color Doppler study (black arrow).

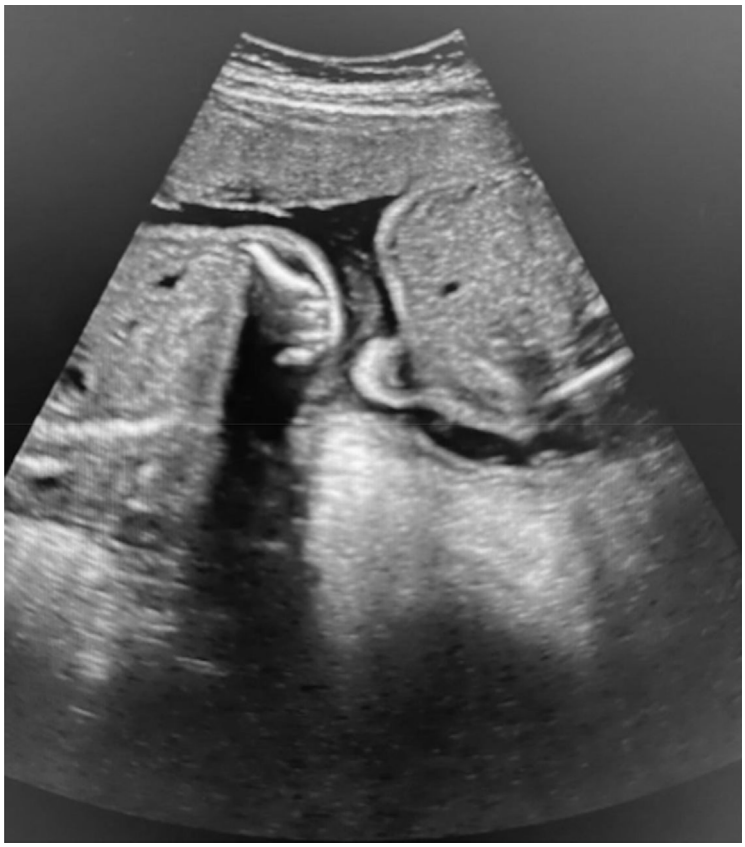

Fig. 2 – Grey-scale US image showing the abdomen of 2 fetuses at 32 weeks of gestation with separating thin membrane in between.

parts. Intake of folic acid during pregnancy is associated with significant decrease in the reduction of congenital anomalies like anencephaly, a serious neural tube defect. This is a lethal condition with no available treatment, potentially resulting in fetal death in utero, during birth, or after birth [7]. With no any history of any medication intake like folic acid during her pregnancy, cause being unspecified, might have contributed to the formation of such anomaly as explained in the previous literature where folic acid deficiency has been proposed to cause neural tube defects like anencephaly [8].

In the presented case, anencephaly was diagnosed at 25 weeks during her first antenatal visit with the help of transabdominal ultrasonography, but it could be diagnosed by transvaginal sonography as early as 11-14 weeks, with the demonstration of high alpha-fetoprotein in serum of the mother during the second trimester. However, in some condi-

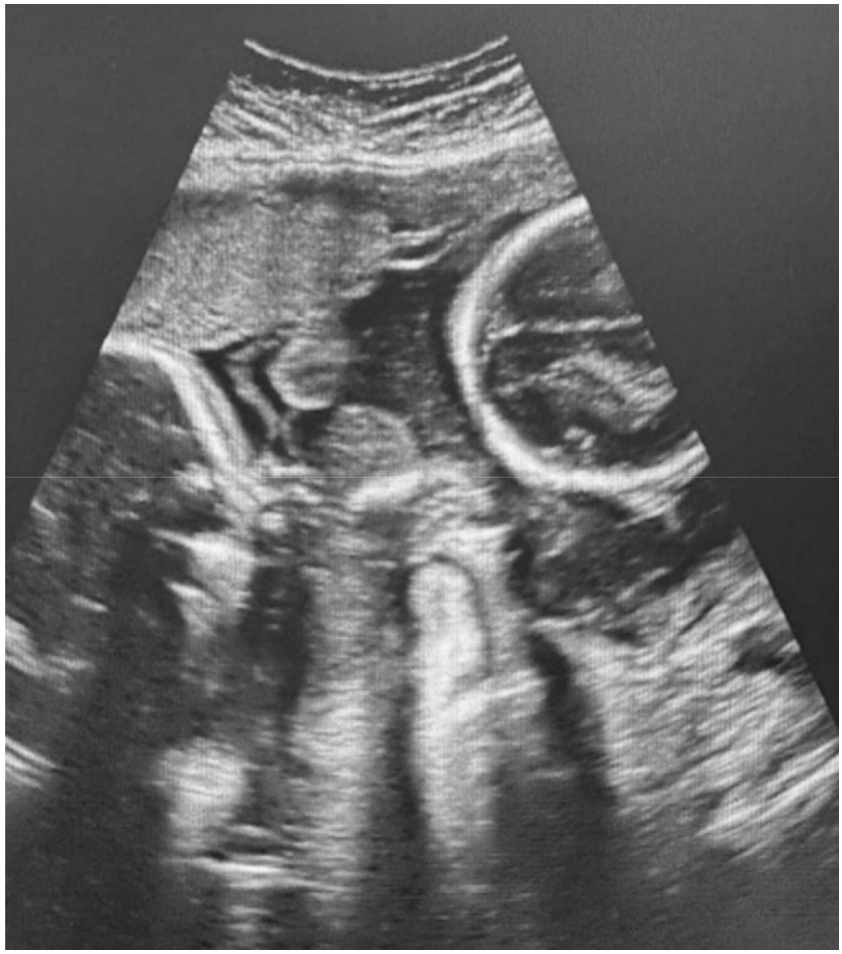

Fig. 3 – Grey-scale US image showing vertex presentation for two fetuses with intervening membrane. The third fetus is not visible.

tions, such as anencephaly, the early ultrasonography features differ from those observed in the second and third trimesters [2,6]. Unfortunately, the patient in the present case lost the follow-up for the subsequent visits which posed a significant challenge in further evaluation and management of the patient.

Anencephaly can be detected through early diagnosis methods such as ultrasound and amniocentesis. In the first-trimester ultrasound, a distinctive "Mickey Mouse sign'' is observed, characterized by 2 semicircular structures resembling Mickey Mouse's round ears above the fetal surface. During the second trimester, a significant reduction in brain tissue results in a "frog face'' or "frog eyes'' appearance, with no visible brain tissue above the eyes. Second-trimester amniocentesis examinations may also reveal elevated maternal serum alpha-fetoprotein levels, which can serve as an indicator of anencephaly. The present case is in accordance with the literature [7,9,10].

It is required to differentiate anencephaly from amniotic band syndrome as the key distinctions include the presence of additional defects (limb or digit amputations, ventral wall defects, spinal defects) not linked to anencephaly, and the asymmetry of the cranial defect in amniotic band syndrome versus the symmetric cranial defect in anencephaly.

In the present case transabdominal ultrasound image showed triplet pregnancy with normal flow in color Doppler study (as shown in the Fig. 1B), also showed triplet pregnancy with 3 separate placentae with absent cranial bone with bulging orbit giving "Frog eye appearance" (as shown in the Fig. 1A). The findings are consistent with the literature findings as mentioned.

The radiological diagnosis relies on identifying the symmetric absence of typically developed brain and skull structures above the eye sockets. Even if angiomatous stroma is present, it should not deter the sonographer from diagnosing anencephaly, nor should its presence be misinterpreted as a factor that could improve the consistently bleak prognosis associated with this condition [10].

Unprecedented spontaneous reabsorption in utero (vanishing triplet)

Vanishing triplet is a fetus in a multi-gestation pregnancy which dies inside the uterus and then partially and completely reabsorbed [11]. It has been postulated that, the incidence of vanishing triplet is due to poorly implanted placenta, and developmental anomalies that may cause major organs to fail or to be missing completely, or there may be a chromosome ab-

normality incompatible with life [12]. Spontaneous reabsorption of an anencephalic fetus in utero is an exceedingly rare and unexpected event, with no documented cases in triplet pregnancies until this report. The mechanisms underlying such resorption remain poorly understood, warranting further investigation. It raises questions about the factors contributing to the resorption of anencephalic fetuses and whether this process differs in multifetal pregnancies. More research is needed to shed light on this unique phenomenon. Spontaneous reabsorption in our case may be due to the presence of documented developmental anomaly (anencephaly).

Unique nature of the triplet pregnancy

This case underscores the uniqueness of triplet pregnancies and the unexpected outcomes they may present. The combination of triplet pregnancy, anencephaly, and spontaneous reabsorption in utero is unprecedented in the medical literature. The management of such complex cases requires close monitoring and individualized care plans. However, the loss of subsequent follow-up posed significant challenges in further managing and evaluating the patient.

Management and delivery

In the present case despite receiving adequate counselling about her triplet pregnancy and fetal reduction at the feto-maternal unit, she chosed to proceed with the pregnancy without reducing the number of fetuses. The majority of anencephalic babies do not survive until birth, often resulting in a miscarriage. Babies born with anencephaly typically have a very short life expectancy, typically measured in days or weeks [3]. There are 2 primary treatment options for anencephaly. The first option is an expectative approach, which involves monitoring the pregnancy without intervention. However, this approach carries a significant risk of hydramnios, particularly when the pregnancy is dichorionic. It necessitates iterative amniotic punctures, which come with potential infectious, serologic, and bleeding risks. The second treatment option is selective feticide, a procedure that entails terminating 1 fetus within a multiple pregnancy after the first trimester. The specific technique used for selective feticide depends on the chorionicity of the pregnancy. Early diagnosis through first-trimester echography is crucial for making an informed decision regarding these treatment options, with the consent of the parents [7,8]. However, presentation in the second trimester and will of the patient to continue the pregnancy despite adequate counselling posed a challenge in management.

After the significant loss of subsequent follow-up, she presented in the third trimester, during the third trimester (32 weeks of gestation) of her pregnancy, seeking medical attention due to symptoms of premature labor. Due to her advanced gestational age and the potential risks associated with a triplet pregnancy, a decision was made to perform a cesarean section (LSCS) to ensure the safety of the remaining 2 viable fetuses. She gave birth to 2 healthy babies with a complete reabsorption of the third anencephalic fetus which was astonishing. This case report presents the rare combinations of triplet pregnancy with an anencephalous fetus undergoing spontaneous resorption.

Conclusion

This case report highlights the exceptional nature of a triplet pregnancy complicated by anencephaly and the unprecedented event of spontaneous in-utero reabsorption of an anencephalic fetus. The management of such cases necessitates specialized care and vigilant monitoring. Further research is needed to better understand the mechanisms behind fetal reabsorption and the unique aspects of multifetal pregnancies, particularly in the context of congenital anomalies. This case serves as a reminder of the importance of thorough prenatal care and the need for ongoing research to improve our understanding of complex pregnancy scenarios.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

REFERENCES

[1] Yıldırım E. Spontaneous triplet pregnancy and trap sequence, case report. BMC Pregnancy Childbirth 2019;19(1):328. doi:10.1186/s12884-019-2484-3.
[2] Fong KW, Toi A, Salem S, Hornberger LK, Chitayat D, Keating SJ, et al. Detection of fetal structural abnormalities with US during early pregnancy. Radiographics 2004;24(1):157–74. doi:10.1148/rg.241035027.
[3] Mikki M. A case report on anencephaly," *Chettinad Heal*. City Med. J. 2022;11(04):92–4. doi:10.24321/2278.2044.202245.
[4] Suleiman WK, Alzahrani AMA, Alomari BAA, Alghamdi BSA, Alghamdi MSS, Alghamdi NAA, et al. Triplet gestation with anencephaly: a case report. Int J Med Dev Ctries 2022;6(11):1413–19. doi:10.24911/ijmdc.51-1664895660.
[5] Chidi E, Sylvester E, Ekop E, Uka O, Odidika U, Dierdre T. Triplet pregnancies in a southeastern Nigerian Hospital: before the artefacts set in,. Trop. J. Obstet. Gynaecol. 2016;33(2):159. doi:10.4103/0189-5117.192217.
[6] Aguemon CT, Lokossou S, Ogoudjobi M, Kamga S, Matabishi B, Diab M, et al. Selective termination of pregnancy: about a case of anencephaly on a bi-chorial twin pregnancy. Open J Obstet Gynecol 2018;08(04):315–20. doi:10.4236/ojog.2018.84034.
[7] Manubulu CCP, Sari IK. Diagnostic problems of pregnancy in a mother with fetal anencephaly. Int. J. Adv. Med. 2023;10(7):581–4. doi:10.18203/2349-3933.ijam20231883.
[8] Joel Momo R. Challenge in the management of twin pregnancy with anencephaly of one fetus in a low- income country: a case presentation. J. Gynecol. Obstet. 2019;7(3):81. doi:10.11648/j.jgo.20190703.15.
[9] Szkodziak P, Krzyżanowski J, Krzyżanowski A, Szkodziak F, Woźniak S, Czuczwar P, et al. The role of the 'beret' sign and other markers in ultrasound diagnostic of the

acrania-exencephaly-anencephaly sequence stages. Arch Gynecol Obstet 2020;302(3):619–28. doi:10.1007/s00404-020-05650-y.
[10] Goldstein RB, Filly RA. Prenatal diagnosis of anencephaly: spectrum of sonographic appearances and distinction from the amniotic band syndrome. Am. J. Roentgenol. 1988;151(3):547–50. doi:10.2214/ajr.151.3.547.
[11] Manzur A, Goldsman MP, Stone SC, Frederick JL, Balmaceda JP, et al. Outcome of triplet pregnancies Dier assisted reproductive techniques: how frequent are the vanishing embryos. Fertil Steril 1995;63:252–7.
[12] Goldman GA, Dicker D, Feldberg D, Ashkenazi J, Yeshaya A, et al. The vanishing fetus. A report of 17 cases of triplets and quadruplets. J Perinat Med 1989;17:157–62.