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Conservative Management of a Twin Pregnancy Complicated with Demise of a Sibling Having Turner Syndrome

Turner Sendromlu İkiz Eşi Kaybı ile Komplike Olan Bir İkiz Gebeliğin Koruyucu Yönetimi

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ABSTRACT: Risk of chromosomal and structural anomalies are increased in twin pregnancies with potential loss of one fetus. Single fetal death in twin pregnancies is believed to be associated with increased risk of perinatal morbidity and mortality for the surviving twin and risk of coagulopathy affecting the mother. In this report we present a case of twin gestation with one of the fetuses having Turner syndrome with a hydrops fetalis and large cystic hygroma compressing the normal fetus. The abnormal fetus demised in the 22nd week of pregnancy. The patient was followed by conservative management and a healthy fetus was delivered by cesarean section. We discuss the risk of consumptive coagulopathy and fetal reduction procedure in a case of twin pregnancy with one demised fetus.

Key Words: Twin pregnancy, Cystic hygroma, Turner syndrome, consumptive coagulopathy.

ÖZET: İkiz gebeliklerde kromozomal ve yapısal anomali riski artmış olup fetal kayıp görülebilir. İkiz gebelikteki tek fetüsün ölümü yaşayan ikiz eşinde perinatal morbidite ve mortaliteyi, annede ise koagülopati riskini artırmaktadır. Bu çalışmada fetüslerden birinin Turner sendromlu olduğu ve fetal hidrops ile büyük kistik higromanın eşlik ettiği bir ikiz gebelik olgusu sunulmaktadır. Anormal fetüs 22. gebelik haftasında kaybedildi ve gebelik koruyucu yönetimle izlenerek sağlıklı fetüs sezaryen ile doğurtuldu. İkiz eşinin kaybının eşlik ettiği ikiz gebeliklerdeki tüketim koagülopatisi ve fetal redüksiyon işlemleri tartışıldı.

Anahtar Kelimeler: İkiz gebelik, Kistik higroma, Turner sendromu, tüketim koagülopatisi

INTRODUCTION

Twin gestations are associated with greater risk of complications and adverse outcome for both mothers and fetuses than singleton pregnancy. Twin pregnancies with discordant anomaly or single intrauterine demise is further complicated and may face a dilemma in management. Turner syndrome is the most common sex chromosome abnormality in females, typically associated with the absence of one sex chromosome (45,X), although mosaicism or structural abnormality in one sex chromosome may also be responsible for the condition. It affects approximately 1 in 2000 liveborn females and it has been estimated that only about 1% of fetuses with Turner syndrome reach to term [1]. The most common prenatal sonographic findings associated with Turner syndrome are cystic hygroma and hydrops fetalis [2]. Cystic hygroma is a subcutaneous accumulation of fluid around the neck or axillary region at various degrees. The incidence of prenatally diagnosed cystic hygroma ranges from 1 in 200 spontaneous abortions to 1 in 600 to 700 low-risk pregnancies [3,4]. Most of the affected fetuses have aneuploidy with Turner syndrome being the most common [5].

In this report, we present the sonographic findings of a dizygotic twin pregnancy in which one fetus had Turner syndrome with huge cervical cystic hygroma and hydrops fetalis. After intrauterin death of the affected fetus in the second trimester, a conservative management strategy with the control of regular coagulation parameters was followed, and the delivery was uneventful with a favorable fetal and maternal outcome.

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CASE REPORT

A 28-year-old female with spontaneous pregnancy after 9 years of secondary infertility was initially admitted to our clinic in the 9th week of pregnancy. A dichorionic diamniotic twin pregnancy was detected sonographically, and the mother was called for regular monthly controls. During routine control at 16th week, a huge multiseptated cyctic hygroma was detected around the neck (Fig. 1). No associated sonographic abnormality was evident. Amniocentesis was performed in the 18th week of gestation and two separate samples of fluid were obtained from both sacs using two different 22 G lumbar puncture needles and the amniotic fluid belonging to fetus with cystic hygroma was labeled. Chromosomal analysis revealed Turner syndrome (45, XO) in the fetus with cystic hygroma. No chromosomal abnormality was detected in the other fetus (46, XX). At the follow-up, fetal hydrops manifested by bilateral pleural effusions, ascites and subcutaneous edema was detected sonographically (Fig. 2), and the size of the cystic hygroma around the neck increased exerting a compression effect upon to the normal fetus. At the 21st week of pregnancy a fetal reduction was offered to the parents to save the normal baby. The family hesitated about it due to religious belief and it was a valuable pregnancy after 9 years of infertility. At the 22nd week of pregnancy the fetal cardiac activity of the fetus with Turner syndrome was lost. Compression onto normal fetus was relieved after the loss of the affected fetus.

Close monitorization for the risk of disseminated intravascular coagulation was performed weekly in the first month after the fetal demise, and biweekly after that. The platelet count, prothrombine, activated partial thromboplastine and bleeding times remained within normal ranges during the rest of pregnancy. In the 39th weeks of pregnancy, a 3600gr of healthy female baby was delivered by cesarian section due to breech presentation. Although the affected fetus was highly masserated, the cyctic hygroma around neck could be identified easily (Fig. 3). The perinatal period was uneventful and the mother and the baby were discharged on the 3rd day postoperatively. The follow-up of the mother and the baby at 2nd and 6th weeks of discharge was normal.

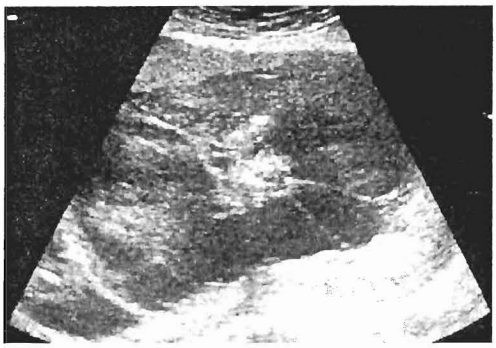

Fig 1. Cervical cystic hygroma surrounding the fetal neck is seen on transverse view.

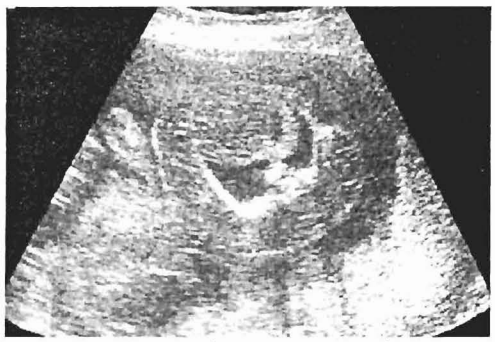

Fig 2. Transverse view through the lower chest and aupper abdomen shows fetal hydrops manifested by bilateral pleural effusion, ascites, and extensive subcutaneous edema.

Fig 3. Picture of the demised fetus with Turner syndrome.

DISCUSSION

Twin pregnancies form 12% of all spontaneous conceptions [6]. Compared to singleton pregnancy twin pregnancies are associated with greater risk of complications and adverse outcome for fetuses and the mothers. The risk factors are mainly determined by the zygocity, chorionicity and number of amnions. There seems to be a benign outcome for the surviving dichorionic twins. On the other hand, the situation may be more dramatic for the monochorionic twins. Therefore, the identification of chorionicity by ultrasound is of great importance for the risk assessment. Twin pregnancies carry a pregnancy loss rate up to 24 weeks of about 6.3% and severe prematurity (24–28 weeks) rate of about 8% [7,8].

This case is interesting because it was further complicated by the intrauterin death of single fetus with Turner syndrome and huge cystic hygroma in the 22nd week of gestation. Chromosomal anomalies and congenital malformations are more common in twin pregnancies compared to singletons. Because each fetus presents an independent risk of aneuploidy, the risk of an affected fetus is approximately doubled in dizygotic twin pregnancy compared to singletons [9,10]. If a fetal anomaly is detected in one of the fetuses, possible management options varies according to gestational age, and the preferences of the physician and the parents. These include 1) termination of the pregnancy, 2) continuation of the pregnancy despite the fetal anomaly, 3) fetal reduction of the abnormal fetus. The type of the chromosomal anomaly was reported to be the main determinant in deciding whether to perform selective fetocide or expectant management in a study investigating the management options in dichorionic twin pregnancies discordant for trisomies [11]. The pregnancy was managed conservatively if the affected fetus unlikely to survive the perinatal period. However, we were unable to find a study exploring the outcome of the dichorionic twin pregnancies discordant for Turner syndrome.

Fetal reduction can be offered in cases of discordant anomalies in twin pregnancies to give a chance of progression of pregnancy for the unaffected fetus. However, selective termination may indeed increase the risk of miscarriage and lead to damage of the co-twin [11]. On the other hand, some anomalies may adversely affect the outcome of the whole pregnancy, or increase the risk of intrauterine death and perinatal morbidity. The overall fetal loss rate due to fetal reduction was reported to be 2.4% with a gestational age of 37.1 weeks at delivery in a series of 164 dizygotic twin pregnancies in which selective termination of anomalous fetus was performed before 24 weeks of gestation [12]. In this case a fetal reduction was offered to the parents after chromosomal abnormality was discovered and to relieve the compression exerted by the huge cystic hygroma over the normal fetus. But because of the religious belief, the family hesitated and the procedure was suspended, and the fetal cardiac activity of the fetus with Turner syndrome was lost spontaneously in the follow up.

Intrauterine fetal demise of one fetus can adversely affect the surviving fetus. Retention of dead fetus or 4-5 weeks in a singleton pregnancy results in an increased risk of maternal consumptive coagulopathy [13]. Single intrauterine fetal death is more common in pregnancies complicated with discordant anomalies [14]. The management of the twin pregnancy with one fetal death varies according to the gestational age. In pregnancies reaching viability a close follow-up and termination of pregnancy after 37 weeks is usually a preferred way of treatment. However, close monitoring is essential in pregnancies at previable stages, because of the risk of consumptive coagulopathy. In twin pregnancies with one demised fetus, the frequency of coagulopaty varies largely among different studies. Although up to 25% coagulopathy rate was reported in one review [15], only one case of maternal consumptive coagulopathy was encountered out of sixteen twin pregnancies [16], and no case of consumptive coagulopathy was detected in 13 twin pregnancies with single fetal death [17]. In our case, the trombocyte count, protrombin and activated partial tromboplastin times, and fibrin degradation products were evaluated weekly as indicators of consumptive coagulopathy. Because all parameters remained within normal limits in the follow up, the pregnancy was terminated safely in the 39th week of gestation with a favorable fetal and maternal outcome.

In conclusion, in case of a twin pregnancy with one demised fetus, a conservative management with close follow up for the signs of consumptive coagulopathy can be chosen for the sake of living fetus until gaining viability. Although contradictory results are present, it looks like the main problem for the surviving fetus is prematurity rather than the siblings death.

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