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Encephalomalacia in surviving twin after single fetal death diagnosed at 18 weeks of gestation in monochorionic twin pregnancy

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEFG Hiroshi Sato
BD Hiroko Murata
BD Kanae Sato
CD Kanako Kawaharamura
CDE Shozo Hamanishi
ADEF Masaya Hirose

Department of Obstetrics and Gynecology, Hyogo Prefectural Tsukaguchi Hospital, Hyogo, Japan

Corresponding Author: Hiroshi Sato, e-mail: shiro801219@yahoo.co.jp

Patient: Female, 28
Final Diagnosis: Single intrauterine fetal death
Symptoms: —
Medication: —
Clinical Procedure: —
Specialty: Obstetrics and Gynecology

Objective: Rare disease

Background: Single fetal death in monochorionic twin pregnancy may result in poor perinatal outcome of the surviving twin, including neurologic sequelae, other organ injury, and death. In most reported cases of poor perinatal outcome in the surviving twin, monochorionic co-twin death occured after more than 20 weeks of gestation, while few with earlier occurrence have been presented.

Case Report: A 28-year-old primigravid woman was referred to our hospital at 18 3/7 weeks of gestation for perinatal management of single fetal death in a monochorionic-diamniotic twin pregnancy. Our first evaluation by ultrasonography revealed a dead twin sized at 16 weeks of gestation, and an alive one with normal size and appearance, together with 1 placenta and 2 amniotic cavities with normal fluid amounts. At 20 3/7 weeks of gestation, ultrasonography showed that the surviving twin had bilateral ventriculomegaly and dilatation all around the subarachnoid cavity despite a normal head size. Fetal magnetic resonance imaging revealed remarkable atrophy of the cerebral cortex. After counseling, the patient and family chose termination of pregnancy, and artificial abortion was performed at 21 weeks of gestation. The aborted fetuses were not anomalous. Autopsy pathological findings confirmed encephalomalacia in the surviving twin.

Conclusions: Recent development of imaging device make it possible that several abnormalities in central nervous system can be detected in detail at earlier gestational age. It is important to keep this condition in mind even though single fetal death occurred at early gestational age.

Key words: monochorionic twin • single intrauterine fetal death • encephalomalacia • fetal magnetic resonance imaging

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Background

Single fetal death in monochorionic twin pregnancy may result in serious complications of the surviving twin, including damage in brain and other organs, and death [1,2]. It has been reported most cases of monochorionic cotwin death with poor perinatal outcome in survivors had fetal death after the late second trimester of pregnancy, while earlier presentation was in few cases [3]. As so the neurologic sequelae in the survivor following monochorionic co-twin death in the first and early second trimester is thought to be rare complication and the accurate frequency is not clear and the clinicopathologic study is not sufficient. Here we report a case of encephalomalacia in the surviving twin following co-twin death diagnosed at 18 weeks of gestation in monochorionic twin pregnancy. We investigated some clinical problems of the neurologic sequelae in the survivor following monochorionic co-twin death in the first and early second trimester.

Case Report

A 28-year-old, gravid 0, para 0 Japanese woman was referred to our hospital at 18 and 3/7 weeks of gestation for perinatal management of single fetal death in a monochorionic-diamniotic twin pregnancy. A previous ultrasound examination performed at 16 weeks of gestation in another hospital showed living monochorionic-diamniotic twins. The fetal size and amniotic fluid had been concordant at that time. There had been no evidence of twin-to-twin transfusion syndrome. Our first evaluation with ultrasonography revealed a dead twin with the size of 16 weeks of gestation, and an alive one with normal size and appearance, together with 1 placenta and 2 amniotic cavities with normal fluid amounts. Maternal vital signs and laboratory data including coagulation function were normal. At 20 and 3/7 weeks of gestation, ultrasonography showed that the surviving twin had bilateral ventriculomegaly and all around dilatation of the subarachnoid cavity despite a normal head size and appropriate interval growth (Figure 1). Doppler study of fetal middle cerebral artery showed the normal peak systolic velocity (22.89 cm/s, 0.9 MoM) [4]. Fetal MRI revealed remarkable atrophy of the cerebral cortex (Figure 2). The patient and her family were informed that brain lesion was likely to be a result of single fetal death in monochorionic twin and that the surviving twin had an remarkably increased risk of neurologic sequelae. They chose termination of pregnancy, and artificial abortion was performed at 21 weeks of gestation. The aborted fetuses were not anomalous. After delivery, blood sample was taken from umbilical vein of the surviving twin. The cord blood hemoglobin concentration was 16.4 g/dl, within normal range. The pathological findings through autopsy confirmed encephalomalacia in the surviving twin (Figure 3). Brain was weighed 13 g, appropriate for 15 to 16 weeks of gestation. Microscopically, neuron density of cerebral cortex was remarkable decreased and migrating neurons were almost absent. Definitive cortical layers were seen but almost injured (Figure 4). In other organs, some infarctions were found at spleen and lung.

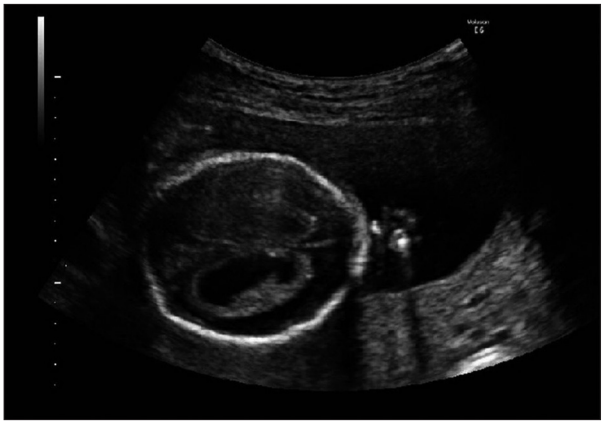

Figure 1. Axial view of the ultrasound scan of the survivor's head at 20 weeks of gestation. This shows bilateral ventriculomegaly and dilatation all around the subarachnoid cavity.

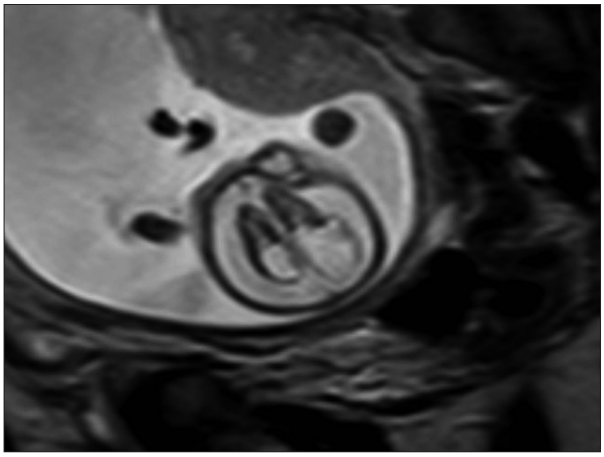

Figure 2. Axial view of the magnetic resonance T2-weighted image of the survivor's head at 20 weeks of gestation. Bilateral ventriculomegaly and dilatation all around the subarachnoid cavity are documented as same as ultrasonography findings. In addition, atrophy of the cerebral cortex is remarkable.

Discussion

Single fetal death in monochorionic twin pregnancy may result in severe brain injury of the survivor. Whereas some theories have been proposed to explain the pathogenesis of the brain lesions found in survivor, including feto-fetal hemorrhage, preceding twin-to-twin transfusion syndrome and congenital

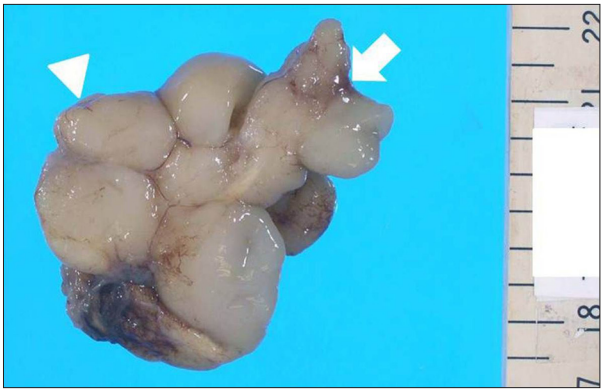

Figure 3. Gross appearance of the survivor's brain (left lateral view). The cerebellum and brain stem were almost normal (arrow). The cerebrum was very softened and atrophic (arrowhead).

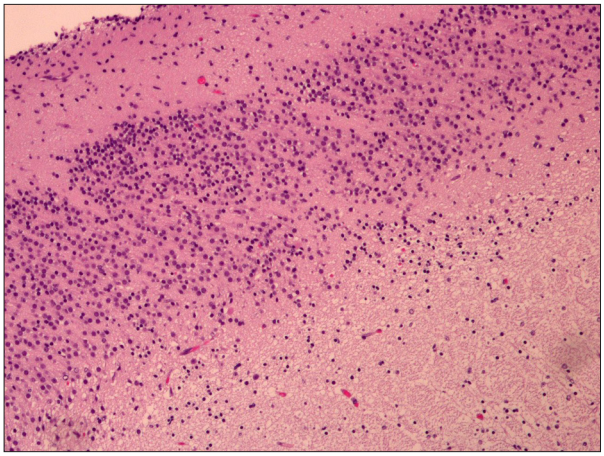

Figure 4. Microscopic findings of the survivor's cerebral cortex. (Hematoxylin and eosin stain). Definitive cortical layers were seen. Neuron density of cerebral cortex was remarkable decreased and migrating neurons were almost absent.

malformations, it is now well established that acute feto-fetal hemorrhage is the main phenomenon involved in the development of brain lesions in survivor [5]. Senat et al. reported that measurement of MCA-PSV was helpful to access anemia of the survivor [6]. In present case, MCA-PSV was not increased and cord blood hemoglobin concentration was normal at termination of pregnancy. But at that time, more than 2 weeks passed after single fetal death, so it is not clear whether survivor was anemic or not immediately after single fetal death. Autopsy pathological study demonstrated encephalomalacia in the survivor, suggesting acute feto-fetal hemorrhage as etiology of brain damage in the survivor. Further investigation concerned in relationship between clinical manifestation, imaging study and pathological evaluation should be warranted.

Relevant English language literature in the Pubmed database was searched and terms used for the database were twin, monochorionic, pregnancy, demise and death in different combinations. Four cases were reported in detail [7–10]. The earliest insult of single fetal death was 12 weeks of gestation [7]. The earliest gestational age of fetal brain damage recognition was is 18 and 6/7 weeks of gestation [10]. Artificial abortion was performed in 1 case. In the other cases, no baby was stillborn but all babies had significant neurologic sequelae. In 2 of 4 cases [8,10], the first ultrasonographic manifestation was ventriculomegaly.

At any gestational age, ventriculomegaly is commonly difined as a measurement of 10mm or greater in the posterior horns of the lateral ventricles noted on an axial brain scan. It is a descriptive term of a pathologic process that has many causes. By using of ultrasonography, we can easily detect ventriculomegaly in early gestational age. In 2 reported cases [8,10] and our case, brain injury with ventriculomegaly tend to be detected earlier only with ultrasonograpy, respectively at 19 weeks, at 18 weeks 6 days and at 20 weeks 3 days. In the other 2 cases [7,9], only microcephaly or head growth restriction was detected by ultrasonography respectively at 28 weeks and at 23 weeks 4 days.

At earlier gestational age, it is more difficult to evaluate fetal central nervous system by using ultrasonography. Recent development of ultrasonography make it possible that several abnormalities in central nervous system can be shown in detail. But it is limited in its ability to visualize the developing cortex and therefore to show cortical malformations [11]. In contrast, fetal MRI can show sonographically occult cortical malformations, as fetal MRI had depicted malformations of cortical development that are sonographically occult, including polymicrogyria [9,12]. Righini et al. [13] and Hoffman et al. [14] also reported that prenatal diffusion weighted MRI detected focal ischemic lesions in the survivor of a monochorionic twin pregnancy within 1–2 days of cotwin death. For early and accurate diagnosis of brain injury, fetal MRI is essential even though when ultrasonography might show almost normal findings. In all reported cases [7–10], MRI was performed and detailed findings were revealed.

The brain grows throughout gestation and early postnatal life. The effect of acute feto-fetal transfusion is thought to be not only destruction of brain, but also interference of development such us differentiation and migration. Autopsy pathological findings show that both of the mechanisms would be occurred in our case.

Conclusions

The frequency of neurologic sequelae in the survivor following monochorionic co-twin death in the first and early second

trimester is not clear. It has been reported that many single monochorionic cotwin death would lead to double death in this period [15]. Therefore, neurologic complications is thought to be rare complication, but some authors have reported this condition. It is important to keep this condition in mind even though single fetal death occurred at early gestational age. When obstetricians manage such patient, they should be well informed about development and anatomy of fetal brain.

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References:

1. Nicolini U, Poblete A: Single intrauterine death in monochorionic twin pregnancies. Ultrasound Obstet Gynecol, 1999;14: 297–301
2. Hillman SC, Morris RK, Kilby MD: Co-twin prognosis after single fetal death: a systematic review and meta-analysis. Obstet Gynecol, 2011; 118: 928–40
3. Bianchi DW, Crombleholme TM, D'Alton ME, Malone FD: Fetology Second edition: Intrauterine Fetal Death In One Twin; New York: Mc Graw Hill Medical, 2010; 810–17
4. Mari G, Deter RL, Carpenter RL et al: Noninvasive diagnosis by Doppler ultrasonography of fetal anemia due to maternal red-cell alloimmunization. Collaborative Group for Doppler Assessment of the Blood Velocity in Anemic Fetuses. N Engl J Med, 2000; 342: 9–14
5. Fusi L, McParland P, Fisk N et al: Acute twin-twin transfusion: a possible mechanism for brain- damaged survivors after intrauterine death of a monochorionic twin. Obstet Gynecol, 1991; 78: 517–20
6. Senat MV, Loizeau S, Couderc S et al: The value of middle cerebral artery peak systolic velocity in the diagnosis of fetal anemia after intrauterine death of one monochorionic twin. Am J Obstet Gynecol, 2003; 189: 1320–24
7. Chen CP, Lin SP, Chiu NC: Microcephaly with dysgenesis of corpus callosum and colpocephaly in the survivor after the first-trimester death of a monochorionic co-twin. Prenat Diagn, 2002; 22: 634–36
8. Weiss JL, Cleary-Goldman J, Tanji K et al: Multicystic encephalomalacia after first trimester intrauterine fetal death in monochorionic twins. Am J Obstet Gynecol, 2004; 190: 563–65
9. Glenn OA, Norton ME, Goldstein RB, Barkovich AJ: Prenatal diagnosis of fetal magnetic resonance imaging in monochorionic cotwin death. J Ultrasound Med, 2005; 24: 711–16
10. Morokuma S, Tsukimori K, Anami A et al: Brain injury of the survivor diagnosed at 18 weeks of gestation after intrauterine demise of the co-twin: A case report. Fetal Diagn Ther, 2008; 23: 146–48
11. Simonazzi G, Segata M, Ghi T et al: Accurate neurosonographic prediction of brain injury in the surviving fetus after the death of a monochorionic cotwin. Ultrasound Obstet Gynecol, 2006; 27: 517–21
12. Jelin AC, Norton ME, Bartha AI et al: Intracranial magnetic resonance imaging findings in the surviving fetus after spontaneous monochorionic cotwin demise. Am J Obstet Gynecol, 2008; 199: 398.e1–5
13. Righini A, Kustermann A, Parazzini C et al: Diffusion-weighted magnetic resonance imaging of acute hypoxic-ischemic lesions in the survivor of a monochorionic twin pregnancy: case report. Ultrasound Obstet Gynecol, 2007; 29: 453–56
14. Hoffmann C, Weisz B, Yinon Y et al: Diffusion MRI Findings in Monochorionic Twin Pregnancies after Intrauterine Fetal Death. Am J Neuroradiol, 2013; 34: 212–16
15. Malinowski W, Koktysz R, Stawerski P: The case of monochorionic twin gestation complicated by intrauterine demise of one fetus in the first trimester. Twin Res Hum Genet, 2005; 8: 262–66