

Twin Pregnancy with Gastroschisis Associated with Sacrococcygeal Teratoma and Limb Abnormalities in both Twins

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Received: February 25, 2020; Published: March 12, 2020

Abstract

Objective: Gastroschisis is a congenital malformation characterized by an abdominal wall defect located laterally to a normal umbilicus. The cause of gastroschisis is unknown, but most probably is considered of exogenous factors. Sacrococcygeal teratoma (SCT) is the most common congenital tumor of the newborn. About 20% of SCTs are malignant. We describe the case of a woman with a twin pregnancy in which both twins had gastroschisis associated with sacrococcygeal teratoma in both twins. A thorough search of literature in PubMed for similar cases did not reveal any reported case of gastroschisis Associated with sacrococcygeal teratoma in both twins.

Case Report: A 21 year old, gravida 2. para 0, abortus 1, woman was referred to fetal medicine unit, Ismailia General Hospital at 22 weeks gestation because of diagnosis of a twin pregnancy with gastroschisis in both twins. Unfortunately, gastroschisis was noted in both twins and another anomalies were observed that were associated with sacrococcygeal teratoma in both twins about 27wks of gestation get spontaneously of preterm labour pain and delivered twins with gastroschisis associated with Sacrococcygeal teratoma. and died within minutes. Both twins presented with a 3-cm abdominal wall defect located to the right side of the umbilicus and a large portion of the bowel protruding that was not covered by a membrane. Histopathology of the placenta revealed that the twins were diamniotic monochorionic. Chromosomal analysis of cord blood showed normal karyotype (46,XX) in both newborns.

Conclusion: Prenatal diagnosis of GS associated with other anomalies should be identified by careful ultrasonographic assessment. Although GS is not usually associated with other anomalies, the present case report makes us aware of the rare incidence of GS with multiple Congenital anomalies and which should be recognized during the prenatal and neonatal periods.

Keywords: Twins Gastroschisis; Sacrococcygeal teratoma

Introduction

Gastroschisis is a congenital defect of the anterior abdominal wall that results in the evisceration of abdominal contents. Gastroschisis is a relatively small defect (2 to 4m) that involves all layers of the ventral wall. The defect is nearly always located just to the right of the umbilicus, although left-side defect have been described in rare instances. The incidence of gastroschisis is 0.94 per 10,000 live births [1].

Sacrococcygeal teratoma (SCT) is a rare subset of germ cell neoplasms but is the most common congenital tumor of the newborn, with an incidence of one in 35,000 - 40,000 live births. SCT is more common in female than male newborn and may occur in twin pregnancies [2,3]. Antenatal diagnosis of SCT can be made by ultrasound during the second trimester or even earlier in the first trimester [4,5].

Citation: Nader Al-labede, Hassan Shora, Fatma Attia., *et al.* "Twin Pregnancy with Gastroschisis Associated with Sacrococcygeal Teratoma and Limb Abnormalities in both Twins". *EC Paediatrics* 9.4 (2020): 34-38.

GS fetuses are also sometimes associated with other anomalies, like congenital hypoplasia or aplasia of limbs due to amniotic bands, pes equinovarus and syndactyly. Teratomas are not usually associated with GS. The overall mortality rate is much lower in isolated GS in conditions when it is not associated with other congenital anomalies. We report a rare case of twins GS fetus showing extracorporeal liver, stomach with sacrococcygeal teratoma. (SCI) malrotated right lower limb and talipes equinovarus of the right lower limb. in both twins.

A thorough search of literature for similar cases in “PubMed” did not reveal any reported twin case of gastroschisis associated with Sacrococcygeal teratoma and limb abnormalities in both twins. To our knowledge this is the first published case of GS associated with SCT and limb abnormalities in both twins.

SCT parasitizes blood supply from the internal and external iliac systems and results in vascular shunting to the rapidly growing tumor. Large tumors during early gestation cause significant prenatal mortality due to placentomegaly, fetal hydrops or high-output cardiac failure [4,5].

Case Report

A stillborn twins GS fetus with SCT delivered in the Department of Obstetrics and Gynecology, Ismailia General Hospital. The mother was 21 years of age, primigravida, at 27 weeks of gestation, and had a history of heaviness in the abdomen and decreased fetal movement for two days. Unfortunately, gastroschisis was noted in both twins, and another anomalies were observed of associated Sacrococcygeal teratoma in both twins also at 22 weeks of gestation get spontaneously of preterm labour pain and delivered twins with gastroschisis associated with Sacrococcygeal teratoma and died within minutes.

She had no abnormal obstetric or medical history. She was non-alcoholic, non-diabetic non hypertensive, and there was no history of serious infection or exposure to teratogens. Ultrasound report at 22 weeks of gestation showed liver and bowel loops freely floating in the amniotic cavity with a midline mass in the gluteal region in both twins. Ultrasound screening performed in Ismailia fetal medicine unit, showed twins with gastroschisis and a cystic mass was observed at the sacrococcygeal region sonography showed a cystic exophytic lower pelvic mass. The mass was skin-covered and septate. It appeared to originate from the tip of an otherwise normal sacrum. Fetal calvaria views were normal, without Chiari II malformation Amniotic fluid average and fetal size was appropriate for gestational age.





The size, location, and cystic composition of the mass and the perceived absence of skeletal dysraphism and Chiari 2 malformation supported the diagnosis of cystic SCT. Fetal MR imaging was recommended to better characterize the mass; however, the patient refused.

Discussion

Gastroschisis (GS) is a major congenital anomaly occurring in the anterior abdominal wall. Usually to the right of the paraumbilical area. The small bowel eviscerates through the defect due to non-rotation and lack of secondary fixation to the posterior abdominal wall. The intestinal loops are exposed to amniotic cavity in intrauterine life and/or exposed to external environment after delivery [6]. The exposed intestines look matted and short because of prolonged contact with hypotonic amniotic fluid particularly during the late gestational period [7]. Prolonged exposure of the intestines to the amniotic fluid may also cause severe complications, such as volvulus atresia, ischemia, increased mucosal permeability and villous atrophy, which lead to the fetal morbidity and mortality in GS fetuses [8]. The GS fetus in the present case showed multiple congenital anomalies, which is very rare. The fetus had evisceration of the whole of the stomach and liver exposed along with bowel loops and was associated with sacrococcygeal teratoma (SCT). The incidence of associated anomalies in gastroschisis is between 10% and 20% and is mostly related to gastrointestinal tract [9]. In 10% of cases, intestinal stenosis or atresia results from vascular insufficiency to the bowel at the time of gastroschisis development. Usually GS is not associated with teratoma, but in the present case it was associated with SCT, which is unusual. The sacrococcygeal region is the most frequent site of teratoma in the fetal and infant stages. In adults, tumors at this site are rare. The incidence of SCT is approximately 1 in 40000 live births and its prevalence is higher in females than in males, with a ratio of 4:1. SCT are germ cell tumors containing the remnants of all three germ layers of the primitive streak, which migrate caudally to rest in the coccyx [10].

SCT are classified into four types: type I is a predominantly external tumor with minimal presacral component and is the most commonly found type in which 85% cases have been observed to be benign tumors. type II is external with a significant intrapelvic component. type III is predominantly internal, both pelvic and intra-abdominal, and with a smaller external component; and type IV is entirely presacral, without an external component or significant intra-abdominal extension. Under this prevailing rule, the SCT found in the present case belongs to type I in which it was completely external [3].

The right lower limb was normally rotated but it showed talipes equinovarus in which the foot was turned inwards, adducted and plantar flexed. This may have been heredity or due to the position of the legs of the fetus within the uterus [11].

Although several possible risk factors have been suggested for the incidence of GS, its exact cause has not been proven. From a developmental basis, the accepted cause for GS is abnormal involution of the right umbilical vein and right omphalomesenteric artery, which leads to ischemic insult to the paraumbilical area through which abdominal organs eviscerate. The other statistically significant risk factors of GS are maternal consumption of alcohol, smoking, use of illegal drugs and young maternal age below 20 years [12].

Prenatal diagnosis of GS associated with other anomalies should be identified by careful ultrasonographic assessment. Although GS is not usually associated with other anomalies, the present case report makes us aware of the rare incidence of GS with multiple congenital anomalies and which should be recognized during the prenatal and neonatal periods.

Nader-Labede syndrome

The definition of syndrome is a set of medical signs and symptoms which are correlated with each other and often associated with a particular disease or disorder [13]. The word derives from the Greek meaning "concurrence" [14]. In some instances, a syndrome is so closely linked with a pathogenesis or cause that the words syndrome, disease, and disorder end up being used interchangeably for them. If an underlying genetic cause is suspected but not known, a condition may be referred to as a genetic association (often just association in context). By definition, an association indicates that the collection of signs and symptoms occurs in combination more frequently than

would be likely by chance alone [15]. Syndromes are often named after the physician or group of physicians that discovered them or initially described the full clinical picture. According this definition and the same multiple congenital anomalies occurred in this case as Gastroschisis and sacrococcygeal teratoma and talipes equinovarus in both twins and there was a case report describe the same concurrent typical multiple congenital anomalies, so giving us new theory as there is a factor that giving us the same concurrent anomalies which are gastroschisis associated with Sacrococcygeal teratoma and limb abnormalities of talipes equinovarus and a new syndrome established named Nader-labede syndrome.

Conclusion

We present the first rare case of Gastroschisis that is associated with sacrococcygeal anomalies of both twins. Chromosomal analysis revealed normal karyotyping 46XX. It is suggested that this case report will increase the awareness of obstetric community of the existence of this rare case report. Further future rigorous researches are needed to understand the sitology, pathogenesis, and innovative strategies of novel approaches to prevent its occurrence.

Bibliography

1. Martin RW. "Screening for fetal abdominal wall defects". *Obstetrics and Gynecology Clinics of North America* 25 (1998): 517-526.
2. Feldman M., et al. "Neonatal Sacrococcygeal teratoma: multi-imaging modality assessment". *Journal of Pediatric Surgery* 25.6 (1990): 675-678.
3. Altman RP, et al. "Sacrococcygeal teratoma: American Academy of Pediatrics surgical survey-1993". *Journal of Pediatric Surgery* 9 (1974): 389-398.
4. Sun DJ, et al. "Early diagnosis of fetal sacrococcygeal teratoma: a case report". *Kaohsiung Journal of Medical Sciences* 19 (2003): 313-316.
5. Roman AS, et al. "First-trimester diagnosis of Sacrococcygeal teratoma: the role of three-dimensional ultrasound". *Obstetrics and Gynecology* 23 (2004): 612-497.
6. Weber IR, et al. "Abdominal wall defects". *Current Opinion in Pediatrics* 14 (2002): 491-497.
7. Sbragia L, et al. "Inflammatory response in a rat model of gastroschisis is associated with an increase of NF kappaB". *Brazilian Journal of Medical and Biological Research* 43 (2010): 160-165.
8. Kale A, et al. "Elevated amniotic fluid amino acid levels in fetuses with gastroschisis". *Brazilian Journal of Medical and Biological Research* 39 (2006): 1021-1025.
9. Molik KA, et al. "Gastroschisis: a plea for risk categorization". *Journal of Pediatric Surgery* 36 (2001): 51-55.
10. Lohfa BC, et al. "Sacrococcygeal teratoma: Clinical characteristics and long-term outcome in Nigerian children". *Annals of African Medicine* 8 (2009): 105-109.
11. Magdalena BS, et al. "Epidemiologic characteristics of amniotic band sequence with Limb malformations without body wall defect: data from the Polish Registry of Congenital malformations". *Archives of Perinatal Medicine* 14 (2008): 23-28.
12. Hui-Fen K, et al. "Twin pregnancy with gastroschisis in both twins". *Taiwanese Journal of Obstetrics and Gynecology* 46.4 (2007): 414-416.
13. The British Medical Association illustrated medical dictionary. Landon: Dorling Kindersley (2002): 177, 536.
14. DorLand's illustrated medical dictionary (32nd edition). Philadelphia, PA: Saunders/Elsevier (2012): 1818.
15. DorLand's Illustrated Medical Dictionary (32nd edition). USA: Elsevier Saunders (2012): 167.

Volume 9 Issue 4 April 2020

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