Fetal intestinal lymphangectasia as a causative factor of mirror syndrome

Dr Amanda Khor
Royal Hospital for Women
Email: khoramanda@gmail.com

Introduction

Mirror syndrome was first described by John Ballantyne in 1892. It is a condition of generalized maternal oedema associated with oedema of the hydropic fetus and placenta.

I present a case of mirror syndrome where fetal ascites is secondary to intestinal lymphangectasia.

Case study

ZA is a 31 year old female, G2P0. She has a past medical history of anxiety and depression. An ultrasound done at 24 weeks gestation revealed gross fetal ascites. The patient developed marked oedema up to the knees and hands at 28 weeks gestation. ZA demonstrated no clinical or biochemical features of preeclampsia throughout her pregnancy.

Maternal serum was negative for cytomegalovirus, herpes simplex, parvovirus and toxoplasmosis.

Fetal echocardiogram showed mild cardiomegaly in keeping with possible high cardiac output. There were no fetal structural abnormalities found on ultrasound. ZA declined NIPT and amniocentesis.

Fetal ascites was drained at 30 weeks gestation. The ascites was tested for syphilis, CMV, parvovirus and toxoplasmosis, all of which were negative. ZA's membranes ruptured a few days after drainage of the fetal ascites and she delivered a baby girl with the assistance of forceps.

ZA's oedema resolved completely after delivery.

At birth, baby of ZA had ascites, skin oedema and chylothorax. The ascitic fluid was chylous. Her 2D echo showed a structurally normal heart. She had normal TORCH serology and faecal Alpha -1 – antitrypsin.

The paediatric gastroenterology team was consulted and diagnosed baby of ZA with a congenital lymphatic malformation or intestinal lymphangectasia.

Discussion

Mirror syndrome has been associated with a variety of causes of fetal hydrops including rhesus isoimmunisation, multiple pregnancies, viral infections, and fetal malformations. Mirror syndrome has been reported to present with pre-eclampsia and/or elevated sFlt1 levels .

A case series demonstrated that treating the cause of fetal hydrops results in resolution of maternal symptoms. In our case, ZA's symptoms only resolved after delivery of the fetus.

Conclusion

Congenital lymphatic malformation can cause fetal ascites and subsequently result in mirror syndrome.

References

1. Braun T, Brauer M, Fuchs I. Mirror syndrome: A systematic review of fetal associated conditions, maternal presentation and perinatal outcome. Fetal Diagn Ther 2010;27(4):191–203.
2. Chimenea A, Garcia-Diaz L, Calderon AM, Moreno-De Las Heras M, Antinolo G. Resolution of maternal Mirror syndrome after succesful fetal intrauterine therapy: a case series. BMC Pregnancy Childbirth 2018 April;18(1):85.
3. Rana S, Venkatesha S, DePaepe M, Chien EK, Paglia M, Karumanchi SA. Cytomegalovirus-induced mirror syndrome associated with elevated levels of circulating antiangiogenic factors. Obstet Gynecol 2007;109(2 Pt2):549–552.
4. Stepan H, Faber R. Elevated sFlt1 level and preeclampsia with parvovirus-induced hydrops. N Engl J Med 2006;354(17):1857–1858.