19th World Congress in Fetal Medicine

A case of spontaneous septostomy in monochorionic diamniotic twins falsely diagnosed as TTTS with fatal consequences for the stuck twin

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Objective

Spontaneous septostomy in monochorionic twins is a rare complication. Only a few cases with low antenatal detection rate have been reported so far.

Methods

We report a case of a monochorionic diamniotic twin pregnancy that presented at 22 weeks of gestation with stage I twin-to-twin transfusion syndrome (TTTS), with polyhydramnios in twin 1 (deepest vertical pocket 12 cm) and anhydramnios in twin 2. Fetoscopic laser photocoagulation was considered, but as both fetuses had visible urinary bladders and normal doppler findings (i. e. no progression to Quintero stage II+), expectant management was preferred. The patient had weekly ultrasound checkups with stable findings and proportional growth (estimated fetal weight difference below 10 %) until week 34, when she delivered by elective cesarean section. During surgery we found that both legs and lower torso of twin 2 were located in the amniotic cavity of twin 1. An amniotic band was attached to his lower abdomen with an apparent strangulation.

Results

After delivery, postpartum adaptation of twin 1 was uncomplicated. Twin 2 was admitted to NICU where cardiorespiratory failure progressed to subsequent death. At autopsy, a deep strangulation was visible in the lower part of the abdomen. No additional malformations were found on the body including the internal organs. A collapsed and almost airless lung, pneumomediastinum and signs of heart failure, ascites and hydropericardium were found. In correlation with the clinical findings, progressive cardiorespiratory insufficiency was concluded to be the cause of death. The etiology of the antepartum rupture of the amniotic membrane is not clear. There was no history of infection, trauma, amniocentesis or other intrauterine procedures. Because the lower abdomen of twin 2 was located in the amniotic cavity of twin 1 and tightly separated, fetus 2 was unable to produce enough fluid to support proper lung development.

Conclusion

Had the septostomy and amniotic band been diagnosed antenatally, amniotic band release by fetoscopy could have been considered to prevent fatal pulmonary hypoplasia. The aim of this case report is to highlight a rare and potentially fatal complication of monochorionic diamniotic pregnancy.