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Prenatal managment of cloacal malformation with lung hypoplasia

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Objective

A 21-year old GI/P0 from Georgia without any pregnancy care had her first ultrasound scan performed at 31 gestational weeks. A complex malformation including dilated intestinal loops, suspicion of duodenal atresia, megacystis and dilatation of the ureter due to suspected atresia of the urethra (prune-belly-syndrome) was detected. A cloacal malformation could not be excluded. As the patient was transferred to refugee accommodation in Bamberg further treatment took place in our prenatal unit in Bamberg.

Methods

This is a case report.

Results

Sonographically a huge abdominal cystic mass reaching from the fetal pelvis to the middle of the abdomen was detected. The content of the mass imposed mildly echogenic. The amount of amniotic fluid was low and the size of the lung small. Duodenum, stomach, testine and heart appeared normal. Due to visible amniotic fluid we concluded a prune-belly-syndrome to be unlikely and suspected the small lung to be related to the huge abdomen elevating the diaphragm to the 4th/5th intercostal space with consecutive compression of the lung. The bladder could not be clearly distinguished from the cystic mass, therefore, a bladder exstrophy could not be certainly excluded. A hydronephrosis (grade 1-2) was found additionally but with normal kidney parenchyma. The sex appeared to be most likely male with hypospadia. An amniocentesis to exclude chromosomal defects was conducted and showed a normal female karyotype (46XX). After maturation of the lung, the cystic structure was punctured (200ml) for diagnostics and to reduce the pressure on the thorax with the intention to improve fetal lung development. The analysis proved the aspirate to be urine. Already one day after the puncture the cystic mass presented similarly again, therefore, in total five punctures were performed over a period of two weeks with a total of 500ml urine to be redrawn. Following the last puncture, rupture of membranes and contractions leading to delivery at 35 weeks occured. The newborn was immediately admitted to the neonatal unit where breathing was sufficient and no severe problems occured. Postpartum findings were a complex urogenital malformation with a total occlusion of the introitus vaginae, clitoromegaly, a persistent sinus urogenitalis, normal anus but rectovaginal communication. Unexpectedly due to an always normally filled stomach, an esophageal atresia with tracheoesophageal fistula (IIIb) was diagnosed which was treated first. At the age of 6 month the child presents healthy. The urogenital malformation is not yet corrected but symptomatically treated with a suprapubic catheter.

Conclusion

A huge cystic mass in the lower abdomen does not necessarily present a lethal condition like prune-belly-syndrome, especially if amniotic fluid is detectable. Due to the prenatal interdisciplinary management the pregnancy could be prolonged for three weeks allowing repeated punctures to achieve better lung function without any need for postnatal assisted ventilation. Therefore, in similar cases punctures at late stages of pregnancy may be reasonable.