Congenital Cystic Malformation of Lungs as a cause of non immune hydrops

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Mrs. AA, 20 years primigravida presented in emergency with antepartum haemorrhage due to placenta praevia and hydramnios at 29^{+5} weeks of gestation. On abdominal examination, uterus was tense and overdistended. Fetal parts could not be palpated and fetal heart sounds were absent. Examination under anaesthesia revealed uneffaced cervix which was only 2 cm dilated, breech presentation and placenta was felt posteriorly reaching upto os. Therefore, an emergency lower segment caesarean section was performed for uncontrolled bleeding and a fresh still-born male baby was delivered. The patient had history of recurrent urinary tract infection for which she had been investigated and treated.

Her haemoglobin was 9.6gm/dl, total leucocyte count and differential counts were normal. Her liver and renal function tests were within normal limits. Urine on microscopic examination showed 10-12 pus cells and Escherichia coli was cultured from urine. Her blood glucose was 76mg/dl and her glucose tolerance test was within normal limits. Total proteins were 6.6 gm/dL with normal albumin/globulin ratio. Her serology for TORCH group of organism and Parvo virus was negative. The couple was non-reactive for VDRL.

An abdominal ultrasound 6 days prior to caesarean section showed live fetus, hydrops fetus with effusion in pleural and pericardial cavities of the foetus along with soft tissue odema. Placenta was posterior and in lower segment, 3 cm from internal os, liquor was increased in quantity.

Complete autopsy of the baby was performed. Baby had protuberant abdomen. Typical faces of hydrops fetalis were not present. There was ascites (100ml), pleural bilateral effusion (20ml each) and pericardial effusion (30ml)). Lungs were solid, unaerated with multiple cysts (measuring 0.5 to 0.8 cm). Other organs did not show any gross pathological abnormality. There was no congenital cardiac abnormality.

Microscopic examination revealed congenital adenomatoid malformation of lungs. There were cystic spaces lined by intestinal epithelium with mucogenic cells (microphotograph 1). In addition, there were alveolar haemorrhages. Placenta measured 30x20x15 cm (weight 320 gm.). No infarct, necrosis or blood clots were present. Microscopically, there were small areas of fibrinoid necrosis along with areas of calcification. All other organs did not show any gross or microscopic abnormality.

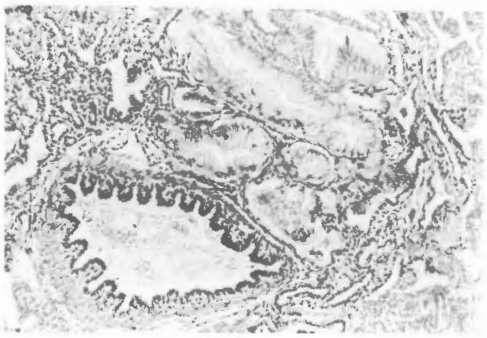

Fig.1 : Photomicrograph showing small cysts lined by mucigenic cells amongst normal alveoli and a bronchiole (H&E, x 100).

Thus, this was a case of non-immune hydrops where the etiology was congenital adenomatoid malformation of lungs. The incidence of non-immune hydrops (NIH) is 1 in 2500 to 1 in 3500 pregnancies but NIH accounts for about 3% of fetal mortality. Congenital cystic adenomatoid malformation of lung is the most frequent lesion in thoracic region, accounting for about 30% cases of thoracic hydrops fetalis. This is one of the ten major fetal diseases (including hydrothorax, chylothorax, laryngeal stenosis/atresia, congenital heart block, sacrococcygeal teratoma, and twin to twin transfusion) amenable to in utero surgery.