Congenital tumors and nonimmune hydrops fetalis

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The sucessful prevention of Rhesus hemolytic disease has resulted in a relative increase in the number of cases of nonimmune hydrops fetalis. During the last year, we observed 10 cases of nonimmune anasarca, of whom five were congenital tumors. Fetal hydrops was due twice to supraventricular tachycardia, twice to adenomatoid lung malformation and once to sacrococcygeal teratoma, neuroblastoma, placental chorangioma, gangliosidosis GM1-type 1, and homozygous alpha thalassemia respectively. In one patient, no cause was found. One of the babies with supraventricular tachycardia conversed in utero by maternal drug administration (Verapamil). Postmenstrual ages ranged from 24 to 40 weeks. All babies showed generalized edema, although only five had birth weights well above the 90th centile for postmenstrual age. Plancental : fetal weight ratios ranged from 0.28 to 0.56 (normal range 0.15 to 0.25), except in one patient with placental : fetal weight ratio of 0.17. In five women, the presenting symptom was hydramnios. Two mothers mentioned no problems, while the remaining three presented with decreased fetal movements, neglected maternal diabetes and maternal palpitations respectively. Ultrasound was performed on 8 occasions and revealed the definite cause of the fetal hydrops in five. Five infants showed a hemoglobin level below 15 g/dl, while two were severely anemic. All albumin levels were below 3 g/dl, except for the conversed infant. Anemia didn't well correlate with the severity of the hydrops, while the hypoalbuminemia (8 patients below 2,4 g/dl) did. Five infants demonstrated marked elevations of the nucleated red blood cell count, while extramedullary hematopoiesis was apparent in 6 out of 8 infants. Generalized edema and hepatomegaly were invariably present. Seven patients suffered from ascites and five from pleural or pericardial effusions. Four babies were stillborn, four died within the first week, and one died at the age of 8 months. The only surviving baby, the one with the conversed cardiac rhythm, develops normally at 4 months of age.

The clinical observations allow some speculations on the etiology of hydrops fetalis. Hypoalbuminemia resulting in decreased colloid osmotic pressure, increased capillary hydrostatic pressure and capillary damage are the key factors. Hypoalbuminemia mainly depends on the decreased synthesis of albumin by the liver and the escape of albumin from the vascular compartment. The increased hydrostatic pressure is secondary to the heart failure, sometimes enhanced by the presence of anemia.

The nonimmune anasarca is a severe condition at birth with poor survival rate, if death has not yet occurred in utero. Early intrauterine diagnosis and appropriate perinatal therapy might improve outcome.

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