A case of Kasabach-Merritt syndrome

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Objective

Kazabach-Merritt syndrome is a rare, potentially fatal complication of rapidly expanding vascular lesions. It is characterized by a combination of hemangiomas, thrombocytopenia and coagulopathy. As an independent nosological form, it is extremely rarely diagnosed prenatally. It is exclusively associated with vascular tumors, hemangiomas, and often accompanies Klippel-Trenaunay-Weber syndrome.

Methods

We present two cases of prenatal diagnosis Kazabach-Merritt syndrome.

Results

Case 1: G1, 25 years old. At 37 weeks, ultrasound examination revealed a fetus with an abnormal appearance and photometric parameters corresponded to the gestational period. The polyhydramnios was determined, the placenta had no ultrasound features, the structure of the umbilical cord was normal, no violations of the fetal-placental blood flow were detected. On examination of the fetal head, in its right part, the formation of a face and neck deforming the outer contour was found. The size of the formation was 11x2,5x15 cm. Study in the 2-D mode had an almost homogeneous medium-echo structure. Multiple diffusely located color loci with low resistance blood flow were recorded in the CDM mode. A large capillary hemangioma of the head was suspected. At 38 weeks, a girl with a mass of 3100 g was born by cesarean section, 47 cm tall, with an Apgar score of 2-3 points. Head circumference - 45 cm, chest circumference - 30 cm. The skin color of the head and neck is purple cyanotic, there was a deformation of the right half of the head and neck due to dense formation, the right auricle was swollen, significantly enlarged in size, purple color. From the first day, the child underwent therapy with prednisone at the rate of 5 μg per kg/mass. With daily monitoring of blood parameters, progressive thrombocytopenia was diagnosed against the background of anemia: at birth, the level of RBC - 2,6x1012/l; Hb - 94 g/l, PLT – 48x109/l. On the 2nd day of life: RBC - 1,6x1012/l; Hb - 44 g/l, PLT - 18x109/l, respectively. A needle biopsy of the neoplasm was carried out: hemangioma. The diagnosis is formulated: Kazabach-Merritt syndrome. The child died on day 5 due to the development of DIC and multiple hemorrhages in the internal organs. Histological examination confirmed capillary hemangioma. Case 2: At 35 weeks, a tumor-like formation was found in the left thigh, deforming its outer contour, 15x12x8 cm in size, polyhydramnios was determined, the placenta did not have ultrasonic features, the umbilical cord was normal, and there were no violations of fetal-placental blood flow. Multiple diffusely located color loci with low resistance blood flow were recorded in the CDM mode. Cavernous hemangioma of the left femur was diagnosed. At 38 weeks a boy with a mass of 3200 g was born by cesarean section, 52 cm tall, with an Apgar score of 6 points. In the region of the lower third of the left femur, knee joint, a tumor-like purple-cyanotic formation was located. From the first day the child was treated with prednisone, platelet mass, fresh frozen plasma. At birth, thrombocytopenia was diagnosed against the background of anemia: RBC - 2.9 × 1012/l; Hb - 85 g/l, PLT - 61 x 109/l. The diagnosis is formulated: Kasabach-Merritt syndrome. Currently, a child aged 2 months to receive therapy with cyclophosphamide and vinblastine with a positive effect. The size of education has decreased.

Conclusion

Prenatal diagnosis of Kasabacha-Merritt syndrome is possible in case of antenatal detection of a large hemangioma, with the development of thrombocytopenia in the neonatal period, anemia of consumption. Neonatal prognosis depends on the severity of hematological disorders.