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PIH as a complication of pregnancy in the patient with rare disease mastocytosis

Katarzyna Stefanska, Marek Niedoszytko, Przemysław Adamski, Dorota Zamkowska and Krzysztof Preis
Medical University of Gdansk, Poland

Arterial hypertension, associated with increased risk of perinatal complications and both maternal and neonatal mortality is diagnosed in 5-10% of pregnant women in Poland. This group includes approximately 5-7% of women with pregnancy induced hypertension, 5-7% of patients with preeclampsia and 1% of individuals with chronic arterial hypertension. Mastocytosis is a myeloproliferative disorders caused by the abnormal proliferation and infiltration of the mast cells in various organs, mainly bone marrow, skin, liver spleen, lymph nodes. The most important clinical symptoms of mastocytosis are related to the symptoms caused by the mast cell mediators release leading to flush, hypotension and in some cases anaphylactic reactions. A 27-year-old primipara at 25 weeks of gestation was admitted to the Obstetrical Department of the Medical University of Gdansk due to PIH. The patient had a 7-year history of systemic mastocytosis suffered from urticaria pigmentosa. Indolent systemic mastocytosis was diagnosed based on 3 minor WHO criteria (tryptase level, D816V KIT mutation, CD2 and CD25 expression on the bone marrow mast cells). She also suffered from grade 3 anaphylactic reactions according to the Mueller scale caused by food and physical factors. The result shows protein loss in urine (5.68 g/day), concomitant decrease in serum concentrations of total protein (59 g/l) and albumins (28 g/l), and an increase of fibrinogen and d-dimer levels. Preeclampsia was diagnosed and treated both pharmacologically and conservatively. The loss of protein was compensated by intravenous infusion of human albumin. FGR was documented on fetal ultrasound. The centralization of fetal circulation was revealed on Doppler ultrasound. Due to preeclampsia, proteinuria and the premises for a possible stillbirth, cesarean section was conducted under spinal anesthesia at 26 weeks of gestation. Early puerperium was complicated by the presence of persistent arterial hypertension and edema. The neonate died at 22 days of life, due to extreme prematurity, circulatory failure and grade 3 intra ventricular hemorrhages. The hypertensive disorders of pregnancy still remain leading causes of maternal and perinatal morbidity and mortality. Mastocytosis is perceived as a medical management dilemma because of its potential for unpredictably heightened mast cell activity in response to various physiologic states including pregnancy. We conclude that proper treatment in pregnant females requires close cooperation of an obstetrician with other medical specialists.

Biography

Katarzyna Stefanska is currently working as a Doctor and Teacher at the Department of Obstetrics of Medical University of Gdansk, Poland. She is interested in perinatology and fetal-maternal medicine, especially in pregnancy induced hypertension-the immunology of PIH, the importance of HLA eplets mother-fetus incompatibility in the pathogenesis of PIH and rare diseases coexisting with PIH-mastocytosis.

kciach@wp.pl

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