Pemphigoid gestationis and intrauterine growth retardation
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Objective
To report a case of pemphigoid gestationis diagnosed in the second trimester and complicated by intrauterine growth retardation. Pemphigoid gestationis (PG) is a rare (incidence: 1/50 000 pregnancies) autoimmune vesiculobullous dermatosis. The disease usually affects multiparous women in the second or third trimester of their pregnancies. Overall fetal mortality does not increase, but miscarriage, preterm delivery, fetal growth retardation and neonatal pemphigoid have been reported as possible complications. These adverse fetal outcomes may be associated with the cross-reaction of the auto-antibodies with collagen XVII in the placenta. Fetal risks are higher in case of an early onset of the disease and in the presence of blister formation.

Methods
This is a case report.

Results
A 34-years-old woman (gravida 2, para 1) was referred due to the high risk (1/219) for Trisomy 21 at the first trimester combined test. The nuchal translucency was within normal limits (NT= 0.9 mm, hCG= 2.09 MoM). At 17 weeks, an elevated maternal serum alpha-fetoprotein (MS-AFP) value of 2.53 MoM was observed. Detailed ultrasound examination at 19 weeks of gestation revealed a male fetus with normal anatomy and biometry consistent with gestational age. However, based on the high risk first trimester screening results, amniocentesis was performed and revealed a normal karyotype. At 21 weeks, the patient presented with severe pruritic rash accompanied by vesiculobuluous lesions located on both upper and lower extremities. The lesions spread rapidly to her abdomen, trunk, buttocks, palms and soles. Face and mucous membranes were not affected. The skin biopsy revealed edema in the papillary dermis, eosinophilic spongiosis, perivascular lymphohistiocytic and eosinophilic infiltration. Direct immunofluorescence showed linear deposition of C3 confirming the diagnosis of PG. As first-line treatment, a topical potent steroid (methylprednisolone aceponate) was initiated, according to the suggestions of the dermatology department, but it failed. Systemic oral corticosteroid (0.5 mg/kg methylprednisolone) and local mupirocin treatment were then chosen. At 25 weeks, all fetal measurements were found to be consistent with 23 weeks gestation. Amniotic fluid index was normal, but umbilical artery Doppler evaluation showed absent end-diastolic flow. A high uterine artery pulsatility index (PI=2.41) with persistent diastolic notch was observed on the placental side, but normal value (PI=1.37) on the non-placental side. Middle cerebral artery and ductus venosus Doppler studies were in normal limits. Asymmetric "mild" ventriculomegaly in the fetal head was also detected, measuring 11 mm on the right side and 9 mm on the left. Corpus callosum and cavum septum pellucidum were normal. Maternal TORCH serology was negative. Fetal MRI confirmed mild asymmetric ventriculomegaly with no concomitant findings. The family was informed about the poor prognosis of early onset- intrauterine growth retardation. The patient's hemogram, biochemical tests and 24-hour urinary protein excretion were normal, inconsistent with preeclampsia. At 27 weeks' of gestation, fetal measurements were concordant with 25 weeks of gestation (abdominal circumference was below third percentile) and accompanied by absent end-diastolic flow in the umbilical artery. A course of intramuscular betamethasone, consisted of two doses (12 mg, 24 hours apart) was administered, with the addition of magnesium sulphate given for 24 hours for fetal neuroprotection. At 29 weeks’ gestation, fetal measurements were consistent with 26+6 weeks and reverse flow in the umbilical artery developed. A male infant weighing 840 g, with respective Apgar scores of 3 and 8 at one and five minutes, was delivered by emergency caesarean section due to fetal distress observed by cardiotocography. There was no skin lesion on the newborn and he was transferred to the neonatal intensive care unit, but unfortunately died on the 11th postnatal day because of the prematurity complications (respiratory distress syndrome, necrotizing enterocolitis, pneumonia, sepsis). The mother was discharged home on the 5th day without any complications in the postpartum period. The steroid treatment was ceased 2 months after the delivery.
Conclusion
In pregnancies complicated with PG, the determinant factors of fetal outcome appear to be the obstetrical factors. Although appropriate pharmacotherapy with a multi-disciplinary approach alleviates symptoms and reduces fetal risks, PG may progress and adversely affect the pregnancy outcome.