AUTOIMMUNE BULLOUS DISEASES

OCCURRENCE OF PEMPHIGUS VULGARIS IN A PREGNANT WOMAN

Dario Didona [1] - Giovanni Di Zenzo [2] - Giacomo Viti [1] - Giovanni Paolino [3] - Biagio Didona [4]

Philipps-university, Department Of Dermatology And Allergology, Marburg, Germany [1] - Istituto Dermopatico Dell'immacolata-irccs, Flmm, Rome, Italy, Molecular And Cell Biology Laboratory, Rome, Italy [2] - Irccs Ospedale San Raffaele Scientific Institute, Unit Of Dermatology And Cosmetology, Milan, Italy [3] - Istituto Dermopatico Dell'immacolata-irccs, Flmm, Department Of Dermatology, Rome, Italy [4]

Background: Pemphigus vulgaris (PV) is a rare autoimmune bullous disease. The onset of PV during pregnancy is a rare event. Indeed, no more than 30 cases of PV arisen during pregnancy have been reported. Newborns of mothers with PV may show PV lesions, because most autoantibodies in PV are IgG4 that can pass through the placenta, leading to neonatal pemphigus vulgaris (NPV).

Observation: A 25-year-old Caucasian pregnant patient was admitted to our Department. She started to complain about pruritus on arms and lower limbs at the 3th month of pregnancy. After one month, she developed flaccid bullous lesions and erosions over the face, scalp, trunk, abdomen, and limbs. Autoantibodies against BP 180 and BP 230 were negative, but antibodies anti-Dsg1 and 3 were 180 and 142 UI/ml respectively. Suprabasal acantholysis and eosinophilic spongiosis have been detected. Direct immunofluorescence showed deposition of IgG on the epithelial cell surface in a smooth pattern. Therefore, a diagnosis of PV was made. She started methylprednisolone 80 mg/die. Unfortunately, the pregnancy resulted in a stillbirth.

Key message: PV in pregnancy has been rarely described. Only 28 cases of PV onset during pregnancy have been described. Indeed, although it has been reported a total number of 61 PV in pregnant patients, among them only 28 were firstly diagnosed during pregnancy. It has been reported that the mean PV onset was during the 16th week of pregnancy.
PV in pregnancy may lead to abortion, fetal growth retardation, intrauterine death, premature delivery, and NPV. NPV is a rare, autoimmune, self-limiting blistering disease.
Treatment options of PV in pregnancy include steroids, steroid-sparing drugs, intravenous immunoglobulin (IVIg), and plasmapheresis. Glucocorticoids are still considered as the first-line treatment. Prednisone is considered the safest drug compared with other glucocorticoids because it does not pass easily through the placenta.