



GENETICS AND GENODERMATOSES

## GYNECOLOGIC AND OBSTETRIC IMPLICATIONS OF DARIER'S DISEASE

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**Background:** Darier disease (DD) has clinical onset in early teenage life (6-20 years' age group) in most of the cases, implying that the disease co-exists with years of reproduction in females. The gynecologic and obstetric issues and complications of DD have rarely been mentioned in the literature.

**Observation:** There are no guidelines for specific management female DD patients. Oromucosal involvement in DD is reported in about 13-50% cases, but female genital mucosal involvement (vagina and cervix) has been described rarely (possibly in no more than two cases). This seems probably due to underreporting. Cytopathologic findings of Papanicolaou smear in DD may lead to erroneous diagnosis of a low-grade squamous intraepithelial lesion. Therefore, the cytopathologist must be informed about the diagnosis for appropriate reporting. Fertility is normal, and pregnancy usually has normal course in DD, but both exacerbations as well as remissions, are reported. Retinoids must be avoided if pregnancy is being planned. Warty papules and plaques of DD can involve groin, vulva and perineum with malodor and pruritus, with potential of secondary infections (e.g., *Staphylococcus aureus*, group B *Streptococcus*). Due to mucosal inelasticity in DD, vaginal delivery may be traumatic and may lead to rupture of the birth canal. Therefore, an elective Cesarean section may have to be planned. In females with lower abdominal lesions, Pfannensteil incision for Cesarean section delivery may be quite problematic. Involvement of skin of back may preclude safe administration of regional anesthesia. A single case report described successful anesthetic management of pregnancy in DD.

**Key messages:** First, for proper interpretation of Papanicolaou smear in DD, cytopathologist must be informed of diagnosis beforehand. Secondly, as DD co-exists with reproductive age group, systemic retinoids should be used very carefully in females. Thirdly, DD may exacerbate as well as improve during pregnancy.

