Pregnancy in Ehlers-danlos Syndrome

A 23-year-old non-diabetic, normotensive primigravida with 36 weeks of gestation was admitted in obstetric inpatients with diminished fetal movements. There was gross intrauterine growth retardation with breech presentation of a living fetus (maturity of 26 weeks with estimated body weight of 1.023 kg), and severe oligohydramnios. The patient had multiple scars in legs which she disclosed has resulted from delayed healing of 'fish-mouth'-like gaping wounds even after minor trauma dating back to her childhood. On examination, the scars were very thin with characteristic 'cigarette-paper' or papyraceous-like, and present over shins, knees (Fig. 1), and forehead. She had depressed bridge of the nose, epicanthic folds and keratoconus. The skin was 'velvety' in feel, very much hyperextensible (Fig. 2) with normal recoil property; neither there was any history of easy bruisability nor were any vessels visible under the skin. The joints were lax and hypermobile (Fig. 3) without any past history of joint dislocation. The patient could extend her tongue to the tip of the nose (Gorlin’s sign) due to increased tissue elasticity. Scoliosis and pes planus (Fig. 4) were present. She was of normal intelligence and without any focal neurodeficit. Examination of the other systems was essentially normal. She was diagnosed clinically as suffering from Ehlers-Danlos syndrome (EDS). Ten clinically distinguishable EDS types have been described in literature. Type IV EDS (vascular type) is most dangerous due to propensity for sudden death from rupture of large blood vessels or hollow organs. Our case fits well with EDS type II (milder form of the classical type I).

Pregnancy outcome in patients with EDS is poorly documented. However, some researchers found that pregnancy is generally well-tolerated in women with Ehlers-Danlos syndrome, with favorable maternal and neonatal outcomes. Commonly encountered problems in EDS with pregnancy are cervical incompetence, recurrent abortions, prematurity, premature rupture of membranes, uterine rupture, ante- and postpartum haemorrhage and tearing of perineal tissue. ‘Amniotic band syndrome’, a very rare entity is classically associated with EDS. This exposes the fetus to fibrous bands from the amnion and chorion which may wrap around portions of the fetus, causing dysplasia or amputation of fetal parts, especially extremities and may even culminate in fetal death. In spite of all efforts, our patient had intrauterine fetal death at 38 weeks of gestation, and she went into spontaneous labor on the next day. A dead male child, weighing 1.250 kg was delivered on the vaginal route. The episiotomy wound was repaired without excessive blood loss. The wound healing was delayed with scar formation.

REFERENCE


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