
CASE REPORT

EHLERS DANLOS SYNDROME WITH PREGNANCY

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A 32 years old lady, G5 P0 A4 was admitted in labour room with gestational amenorrhoea of 20 weeks with the history of fall on the same day, as an emergency case. She was fully conscious when received and her vitals were stable, except the increased respiratory rate. Abdomen was soft, consistent with the period of gestation, no marks of external injury were noted on the body, except 3 bruises over her arm and forearms. On investigation, her blood group was found to be A positive, Hb 10.5 g/dl and platelet count 182,000/mm³. Rest of biochemical investigations were normal. Sonographic finding showed single active fetus of 20 weeks of fetal biometry. There was an episode of bleeding vaginally, which was mild. She had been seen by cardiologist for increased respiratory rate. Her echocardiography was normal, but she was given tablet lasoride and digoxin. Opinion of a consultant physician was asked. After thorough history and examination, she was diagnosed as a case of Ehlers Danlos syndrome, on the basis of hypermobile joints, high arched palate, bruises due to fragile blood vessels, proximal muscle weakness. The final management plan was to build up her Hb%, omit digoxin and lasoride, having one pint of blood in hand, termination of pregnancy was planned after counseling the patient.

The blood was arranged and at 26 weeks of gestation, induction was initiated with prostaglandins. She did not start any uterine contraction, it was repeated again and after the failure of establishment of uterine contraction, her caesarean section was planned.

It was performed in the presence of senior anaesthetist. She delivered a baby girl of 0/10 A/S weighing 1.5 kg. Bilateral tubal ligation was performed. She did well in her postoperative period, except some shortness of breath which resolved spontaneously.

The Ehlers Danlos syndrome is a group of genetic disorders that have resulted from defects in a collagen molecule which would give strength and adhesion to the body tissues.¹ Common features include easy bruising, joint hypermobility, skin that stretches easily, and weakness of tissue.¹ It is categorized according to the form of genetic transmission into different types with many features differing between patients in any given type. In 2001, re-

searches discovered a new form of Ehlers Danlos syndrome that is caused by an inherited abnormality in a protein other than collagen that also normally plays a role in binding together the cells of our tissue (including the skin, tendons, muscle and blood vessels). Abnormalities in this protein called tenascin, also showed that tenascin could play a role in regulating the normal distribution of collagen in connective tissues of the body.²

The different types are, classical type, hypermobility type, vascular type, kyphoscoliosis type, dermatosparaxis type and lastly tenascin X deficient type.³ It is believed to affect 1 in 5000 people. The healing of both accidental and surgical wound is frequently very poor leaving dramatic scar, other problems often include prolapse of mitral valve, aneurysms, hernia and a range of orthopedic and dental problems. The manner in which it affects patient in degree of severity is entirely unpredictable. A Dutch study was conducted in pregnancy with Ehlers Danlos syndrome. The conclusion of that study was pregnancy well tolerated in a woman with Ehlers Danlos syndrome, with favourable maternal and neonatal outcome⁴. In Ehlers Danlos syndrome type-IV it may be associated with severe maternal complication. Pre-conceptual counseling concerns specific possible complication and multidisciplinary approach are recommended⁵. The study was conducted by Department of Obstetrics and Gynaecology, Weslende Hospital, to assess the course and outcome of pregnancies with Ehlers Danlos syndrome with the aim of developing guidelines for assessment of risk and counseling and for providing optimum medical and obstetrical care. The Ehlers Danlos syndrome is treated according to what particular manifestations present in a given individual. Skin protection is critical. Wound must be treated with great care and infections treated and prevented. Joint injury must be avoided; contact sports and activities involving joints impact should be avoided.

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