**Título:** Loeys-dietz syndrome and pregnancy – a case report

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**Área Terapêutica/Tema:** Anestesia Obstétrica (Obstetric Anaesthesia)

**(TEM FOTO)**

**Resumo:**

Background: Loeys-Dietz syndrome (LDS) is a rare autosomal dominant disease characterized by cardiovascular findings like aortic root aneurysms, arterial tortuosity, patent ductus arteriosus and atrial septal defects. Skeletal changes as scoliosis, cervical spine deformity and instability and tissue findings as dural ectasia can be present as well. Individuals with LDS are predisposed to widespread arterial aneurysms and pregnancy-related complications including uterine rupture and death, constituting a challenge to the anesthesiologist. We present a case report of a pregnant woman with LDS surveilled at a tertiary high-risk hospital.

Case report: Primiparous woman, 37 weeks of gestation, 29 years old, LSD and diabetes type I. Body mass index of 25 Kg/m2 with no difficult airway prediction.  Multidisciplinary counseling was provided in this institution. A c-section under general anesthesia with rapid sequence induction was planned. In the event of a massive hemorrhage, a strategy was delineated with the imuno-hemotherapy department. Besides ASA standard monitorization, invasive arterial pressure and BIS were used. Hemodynamic stability throughout laryngoscopy and surgery was assured with administration of opioids pre induction. The delivery was performed without major complications, with 500ml of blood loss. Uterine contraction was assured with oxytocin. Post-operative pain management and nausea and vomiting prophylaxis was planned in advance

Discussion: Neuraxial anesthesia is the approach of choice in elective c-section. However, in LDS patients its pros and cons should be carefully weighted and discussed with the pregnant. The patient ought to be submitted to lumbar magnetic resonance with contrast if neuraxial anesthesia is to be performed, but the possible adverse effects of contrast agents in the fetus cannot be ignored. The existence of dural ectasia, could lead to dural puncture during epidural catheter placement or unsuccessful spinal anesthesia. In the event of neuraxial anesthesia, spinal block should be preferred. In this patient, magnet resonance wasn’t performed and to minimize the potential risks associated to this technique general anesthesia was the chosen approach. Anticipating the possibility of major hemorrhage, every step expected for a massive hemorrhage protocol were considered prior to the c-section, namely blood samples with fibrinogen dosing, blood units available within a short period of time and two vascular accesses. In order to avoid sudden hemodynamic changes that could lead to major hemorrhage, the patient was monitored with arterial line. Reference to intensive care unit was considered.

Learning points: Although LDS is a rare disease, it is associated with many life-threatening medical features. Anesthetic management adapted to the disease is paramount in ensuring successful surgical outcome and to limit morbidity and mortality.

Refrences:

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