KLIPPEL-TRENAUNAY SYNDROME IN ANESTHESIOLOGY

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BACKGROUND

Klippel-Trenaunay Syndrome (KTS) is an infrequent non-hereditary congenital malformation caused by mutations in PIK3CA gene, which causes soft and bone tissue hypertrophy affecting lower limbs, with cutaneous hemangiomas and varicosities. Epidural and intramedullary space can be affected, as well as pelvic organs and birth canal. Hemorrhagic complications are observed due to alterations in hemostasis and coagulation, presenting many challenges to the anesthesiology during labor.

CASE REPORT

30-year-old primiparous woman, 38+3 gestation weeks, diagnosed with KTS. Left leg hypertrophy. A few years ago, femur elongation with skin grafting was performed due to malleolar ulcer, with general anesthesia and no incidences. Nowadays, patient was having medical follow ups with no treatment.

Patient presented worsening of her condition with lumbar pain and paresthesia in her lower limb. Physical examination and echography ruled out vascular malformations in gynecological area, and labor was scheduled due to worsening of her disease.

MR images were studied, and arteriovenous fistulae were found at the cauda equina level and after assessing benefit/risk balance, we decided not to perform epidural anesthesia for labor.

Cesarean section was indicated due to failure to progress.

In the OR, arterial pressure, frequency and oxygen saturation was monitored and laboratory tests were checked, with normal coagulation results. General anesthesia was performed with fentanyl, Propofol and rocuronium. The baby presented Apgar of 9 and 10 after first and fifth minute, birth weight of 2520 gr.

There were no complications such as massive hemorrhage in the perioperative period and no intrabdominal vascular malformations were observed

DISCUSSION

KTS may be associated with vascular malformations in the CNS, as well as hemostasis alterations, that may contraindicate locoregional anesthesia.

Presence of arteriovenous fistulae in cauda equina ruled out axial anesthesia in our patient.

Nowadays, this kind of anesthesia is acceptor for these patients, if vascular spinal malformations had been ruled out by imaging techniques. This is the reason why an interdisciplinary managing before labor are needed to know the extension of the disease and to perform and individualized anesthesia.

It is necessary to rule out vascular malformations