ANESTHETIC MANAGEMENT OF A PREGNANT PATIENT WITH FABRY'S DISEASE.

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Background:

- ❖ Fabry's disease (FD) is a rare systemic X-linked disease.
- ❖ It is related to a glycosphingolipid metabolism disorder in the alpha-galactosidase A enzyme.
- ❖ This results in anatomic abnormalities and organ dysfunction that can increase the risk of anesthesia complications.
- ❖ Epidural block is debated in patients with neurological diseases and recommendations are not unequivocal.

Case Report:

27 y.o. ♀ in week 38 of pregnancy with FD.

Clinical symptoms:

- ✓ fatigue
- ✓ irritable bowel
- ✓ fingers, hands and legs pain
- ✓ hearing loss
- ✓ dyslexia and migraine
- ✓ no treatment.

Preoperative evaluation:

- ✓ ASA II
- ✓ BMI 34
- ✓ proteinuria (667 mg/24 hs)
- ✓ important oedema in face and lower limbs

Difficult airway and ventilation predictors:

- ✓ Mallampati III
- ✓ interincisor distance < 3 cm
- ✓ poor cervical mobility
- ✓ bite test grade II
- ✓ obesity.

- Epidural catheter for labour pain relief

- C-section for non-progressive labour.
- Anaesthesia was provided via the epidural catheter.
- There were no complications.

Discussion :

- ❖ General anaesthesia must be avoided when possible for obstetric patients, due to difficult airway risk among other issues.
- ❖ Furthermore, cases of FD submitted to other interventions usually require the use of video laryngoscopes or supraglottic devices.

Learning points:

- ❖ An exhaustive preoperative evaluation is paramount to design the anaesthetic plan.
- ❖ Epidural anaesthesia was used with success without any change in the clinical status.
- ❖ To our knowledge, this is the first report of neuro-axial regional anaesthesia performed in a parturient with FD.

References:

Anesth Analg 2017;125:822–36; Anesth Analg. 2017 Sep;125(3):716-718.; Bosn J Basic Med Sci. 2017 Jun 6.