Pseudoxanthoma Elasticum in Pregnancy - Anesthetic and surgical considerations -

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Background

Pseudoxanthoma Elasticum (PXE) is a rare inherited connective tissue disease characterized by degenerative changes due to calcification and fragmentation of elastic fibers on the skin, eyes and cardiovascular system. [1]

There are very few case reports in literature describing labour analgesia (LA) in women with PXA. Women with this disease are advised against becoming pregnant, given the concurrence of the risks inherent in this condition with the physiological changes of pregnancy. [1,2]

The aim of this report is to describe the analgesic approach performed in this particular case and discuss the main anaesthetic and surgical considerations in parturients with PXE.

Legend:

Fig. A: Photograph of the axilla showing several yellowish waxy papules;

Fig. B: Fundus of the eye showing angiod streaks characteristic of PXE;

Fig. C: Histological examination of the skin biopsy showing fragmentation of elastic fibres in the upper reticular dermis. (HE, 100x) [3]

Case report

37 year-old parturient, ASA II, with the following background:

- PXE (diagnosed at age 18 by skin biopsy);
- Dyslipidemia and obesity;
- IIIGOP with known history of infertility and previous assisted reproduction techniques which turned into two pregnancies, both followed by abortion complicated with organ dysfunction and need for ICU hospitalization.

She was admitted at the Emergency Department for threatened preterm delivery at 33w+6d. The present gestation resulted from a spontaneous pregnancy without registered complications, aside elective cerclage at 17w.

Objective examination showed yellowish spots on the flexures and fibrotic streaks in the retina, both PXE features. Cardiomegaly was noted on chest x-ray.

Our Approach...

Based on the literature and given the parturient's pathology, we proposed a Locoregional Technique (LRT): epidural and normal delivery, that the patient accepted.

Epidural was performed uneventfully and labor time was reduced through the use of forceps (♀ live birth, 2250g, Apgar 8/8). During placental removal, the umbilical cord ruptured, which led us to the operating room for manual internal removal under increased safety and monitoring. This procedure was performed under an epidural anaesthetic bolus and light sedation allowing a **good control of vital signs, as desired**.

In the immediate postpartum period, there was no record of obstetrical or anaesthetic complication. The puerpera was discharged after four days, clinically stable.

About pregnant women with PXE:

- ✓ Necrotic and placenta mineralization abnormalities are common, which can cause fragilities and accidental ruptures.
- ✓ Monitoring and control of heart rate is crucial due to premature development of atherosclerosis and risk of acute myocardial infarction.
- ✓ LRT allows for greater haemodynamic stability, with less likelihood of bleeding from injured arteries and reduced cardiac work during labor.
- ✓ There may be bleeding from the retina secondary to the Valsalva maneuvers during the expulsive stage of labour. Therefore, an instrumented delivery is recommended to shorten its duration.

We present our successful case using a LRT to recall the approach of LA in pregnant woman with PXE and to aid in the decision process for the most appropriate analgesic choice, taking into account the few cases described and anesthetic implications still being poorly known.

References:

- [1] *Int J Obstet Anesth.* 2003; 12(1):45-7
- [2] *Rev Cubana Obstet Ginecol.* 2007; 33(2)
- [3] *Med J Aust.* 2014; 201(9):544