

Anesthetic Management of Labor in a Woman with Hereditary Angioedema

Gestão Anestésica de uma Mulher Grávida com Angioedema Hereditário

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Palavras-chave: Analgesia Obstétrica; Anestesia Obstétrica; Angioedemas Hereditários

Hereditary angioedema (HAE) is a rare genetic disorder marked by recurrent swelling episodes in the skin and mucous membranes due to C1 inhibitor (C1-INH) deficiency or malfunction, leading to bradykinin overproduction.^{1,2} This condition poses significant challenges during pregnancy,² and requires proper anesthetic management during labor.

We describe a case of a 30-year-old primigravida with HAE and normal C1-INH levels who experienced recurrent angioedema episodes since a young age, occasionally affecting the glottis, and which responded to frozen fresh plasma. Multidisciplinary collaboration was essential, requiring delivery at a facility with plasma-derived human C1-INH concentrate (PdhC1INH). During pregnancy, despite having discontinued treatment with danazol, the frequency of angioedema decreased, with only mild episodes in the third trimester.

At 38 weeks and four days, she underwent induction of labor, receiving short-term prophylaxis with PdhC1INH infusion one hour before the epidural catheter placement. An experienced anesthesiologist used ultrasound to locate the cricothyroid membrane. A vacuum-assisted delivery was performed, resulting in a healthy newborn. The patient was monitored in the high-dependency unit (HDU) for 24 hours and discharged after two uneventful days.

This case illustrates the successful management of HAE during labor and postpartum. HAE, affecting approximately 1 in 50 000 individuals worldwide,¹ involves recurrent edema formation in various body sites. Types 1 and 2 involve C1-INH deficiency or dysfunction, respectively, while normal C1-INH levels characterize type 3, posing challenges during pregnancy due to estrogen susceptibility.³

Plasma-derived human C1-INH concentrate is preferred for acute treatment and short or long-term prophylaxis during pregnancy and labor.¹ According to the literature, routine prophylaxis before uncomplicated vaginal deliveries is not recommended, but otherwise advised before forceps or vacuum extraction, or cesarean delivery.^{1,4} The multidisciplinary team decided to administer PdhC1INH since

the patient had angioedema crises during the third trimester,¹ no long-term prophylaxis was administered during the pregnancy, and had a known history of multiple laryngeal attacks.⁴ Epidural analgesia might avoid airway complications, especially during cesarean delivery.¹ Prompt identification of potential airway compromise is crucial, considering the risk of laryngeal edema, since it may not respond to standard treatments.^{4,5} The postpartum period requires close monitoring in the HDU due to potential crises in the first 72 hours, which can be treated with PdhC1INH when necessary.⁴

Effective management of pregnant women with HAE requires a multidisciplinary approach, individualized labor plans, and proactive measures to address potential complications. Prophylactic PdhC1INH and epidural analgesia are pivotal in preventing HAE attacks during labor and delivery, thus ensuring favorable outcomes.

AUTHOR CONTRIBUTIONS

IPR: Study design, data acquisition and analysis.

MMA, LM: Study design, critical review of the manuscript.

All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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