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CLINICAL INFORMATION

Anesthetic considerations for a patient with hereditary angioedema – A clinical case

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Abstract Hereditary angioedema (HAE), with an estimated prevalence of 1:50,000, is a rare but potentially fatal disease. It may present with recurrent systemic edema of the subcutaneous tissue and mucous membranes. Patients with HAE are at increased risk for clinical worsening with surgical stress, and may develop respiratory distress syndrome due to impaired airway and hemodynamic instability. The perioperative management of these patients requires specific interventions. We present a clinical case of a woman, 50 years old, with HAE type II scheduled for **ureteral stent placement** via endoscopic approach.

PALAVRAS-CHAVE
Angioedema hereditário;
Imuno-hemoterapia;
Profilaxia

Considerações anestésicas perante um doente com angioedema hereditário – Caso clínico

Resumo O angioedema hereditário (AEH), com uma prevalência estimada de 1:50000 pessoas, é uma doença rara mas potencialmente fatal. Pode se apresentar com edema sistêmico recorrente do tecido subcutâneo e das mucosas. Os doentes com AEH têm um risco acrescido de agudização clínica com o estresse cirúrgico, podem desenvolver síndromes de dificuldade respiratória por compromisso da via aérea e de instabilidade hemodinâmica. A abordagem perioperatória desses doentes requer intervenções específicas. Apresentamos um caso clínico de uma mulher de 50 anos com AEH tipo II indicada para realizar ureteroscopia com colocação de *stent*.

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Introduction

Hereditary angioedema (HAE) is an autosomal dominant disease characterized by quantitative or qualitative changes in the level of the first complement cascade component inhibitor (or C1 esterase inhibitor – C1-INH), allowing uncontrolled activation of the classical complement cascade.[1,2] HAE classification had two variants phenotypes: type I is characterized by low plasma levels of normally functional C1-INH; type II is characterized by the presence of normal or high plasma levels of C1-INH nonfunctional or abnormally functional.[1] More recently, the existence of a third type of an estrogen-dependant HAE with normal functional and quantitative levels of C1-INH has been identified.[3]

Despite its name, C1-INH not only acts on the complement cascade, it also inhibits proteases from coagulation/fibrinolysis cascade and kinin pathways. In HAE, the associated symptoms are mainly due to interference on kinin pathways with increased bradykinin production. Clinically, HAE may present with systemic and recurrent edema of subcutaneous tissue or mucous membrane, with involvement of the gastrointestinal system. It can be misinterpreted as an acute abdomen condition; airway condition with respiratory distress, laryngospasm, bronchospasm, and choking; hemodynamic instability due to anaphylactic shock and death.[1,4] Trauma is the precipitating factor of the clinical picture in about one third of patients. Other triggering factors are infection, anxiety, and estrogens. In a significant number of cases, the precipitating factor is not identified.[5,6]

Case report

We describe the perioperative anesthesia care to a 50 years old woman scheduled for urologic elective surgery: ureteroscopy with stenting for non-radiopaque kidney stones. The patient had a diagnosis of HAE type II and a positive family history (father and brother with the same condition). She was being followed-up at the hospital with immunoallergology consultation and had no regular therapy.

After careful preanesthetic evaluation, and in accordance with the preoperative protocol of the immune-hematology service of our hospital and her immunoallergology consultation, prophylaxis was performed with four oral doses of stanozolol 4 mg (Winstrol®) two days prior to surgery and administration of 1000 U of C1-INH concentrate (Berinert®) 45 min before surgery, at slow intravenous bolus. Premedication with diazepam 1 mg was given at the night before surgery and on the morning of surgery.

On the day of surgery and upon arrival in the operating room, she was monitored with standard monitoring and pretreated with intravenous midazolam (2.5 mg). Subarachnoid blockade was the anesthetic technique of choice, performed with a 25G beveled needle at L3–L4 level with bupivacaine 0.5% (15 mg), with median approach and a blockade up to T10. Parecoxib (40 mg) was administered and antibiotic prophylaxis performed with intravenous cefoxitin (2 g). According to the protocol, 500 U of C1-INH concentrate (Berinert®) were maintained on standby in the operating room, in the event of an acute clinical picture.

The surgery lasted 20 min; the patient remained hemodynamically stable during surgery and in the immediate postoperative period. Postoperative care did not require specificity associated with HAE, and the patient was treated with intravenous paracetamol and IV parecoxib, antibiotic and fluid therapy. The patient was discharged after 24 h.

Discussion

There are numerous factors precipitating acute exacerbations of HAE, particularly perioperative anxiety and surgical trauma, hence the importance of an adequate premedication.

The acute episode approach, with symptoms and varying severity, is controversial. Evidence-based medicine suggests that acute exacerbations may not respond to treatment with epinephrine, antihistamines, or glucocorticoids, a more specific therapy is required.[5,7] The initial approach of a severe acute episode should include the use of recombinant C1-inhibitor drugs or bradykinin receptor antagonists.[2,3,6,7] When these drugs are not available, other approaches include the use of higher doses of androgens, such as danazol or derivatives, tranexamic acid (TA), epinephrine (not always effectively), analgesic control, fluid therapy, and intensive supportive therapy.[5,6] The use of fresh frozen plasma remains controversial due to its theoretical potential to exacerbate and perpetuate the shock.[6]

The most appropriate approach for this pathology is the prophylaxis.[5,6] Thus, in elective surgery, the approach should include anxiolytic premedication and:

(a) Use of recombinant factor C1-INH in the following dosages: 500 U (if <50 kg); 1000 U (if >50 kg, but <100 kg) or 4000 U (if >100 kg) 30–60 min prior to the surgical procedure, with daily repetition if the risk of precipitating an acute episode remains high.
(b) Androgen derivatives, up to five to seven days before surgery.
(c) Eventually TA, although not as effective as androgen derivatives.
(d) Double prophylactic therapy, as was our option.

The regional anesthesia technique option also presents advantages over general anesthesia, as it does not imply an airway active manipulation that, by triggering an episode of exacerbation with a more localized impact, could lead to a laryngeal edema.

In conclusion, HAE is a rare disease, with a few contact by most anesthesiologists. Due to the potential for originating severe complications, it requires a careful perioperative preparation, with the involvement of multidisciplinary teams (immuno-hemotherapists, surgeons, critical care physicians, immunoallergologists, and anesthesiologists), adoption of an appropriate prophylaxis, and correct monitoring of the patient.

Conflicts of interest

The authors declare no conflicts of interest.

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