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Case Report

Perioperative Management of a Patient with Hereditary Angioedema undergoing Lumbar Interbody Arthrodesis

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Abstract

Hereditary angioedema (HAE) is a rare genetic disorder resulting from an inherited deficiency or dysfunction of the C1-esterase inhibitor. It is characterized by recurrent and self-limiting angioedema episodes, most often affecting the skin or the mucosal tissues of the upper respiratory and gastrointestinal tracts.

Androgens and fresh frozen plasma (FFP) are frequently used as treatment despite multiple, significant side effects. Newer therapies include C1-inhibitor concentrate (C1-INH) and contact system modulators such as ecallantide and icatibant. In addition, C1-INH can also be used for prophylaxis.

We present the case of a 47-year-old male with a previous diagnosis of HAE, underwent elective lumbar interbody arthrodesis under general anesthesia. Following prophylaxis with danazol and C1-INH, surgery was carried out uneventfully with an unremarkable postoperative course. Perioperative prophylaxis to avoid acute attacks and anesthetic management are discussed.

INTRODUCTION

Hereditary angioedema (HAE) is a rare genetic disorder caused by a deficiency of C1-inhibitor. These patients suffer acute episodes of angioedema, involving any area of skin, upper airway, or abdomen .Facial edema may occasionally progress to laryngeal swelling, which can be life threatening, causing prolonged intensive respiratory care or even death from asphyxia.

Perioperative prophylaxis with danazol and C1-inhibitor concentrate (C1-INH) are commonly used to prevent angioedema episodes. Anesthesia management and perioperative implications have been recently commented by some authors [1,2]. We present a patient who had previously diagnosed HAE who underwent successful elective lumbar interbody arthrodesis under general anesthesia. Perioperative measures to prevent acute edema attacks and anesthetic implications are discussed.

CASE REPORT

We present the case of a 47-year-old male with a previous diagnosis of HAE. Over the last 14 years, he presented some episodes of angioedema affecting hands, arms and feet with erythematosus and urticarial lesions. Laboratory data were: C1-inhibitor = 0.10 g/L (normal range = 0.19 - 0.34 g/L) and C4 = 0.12 g/L (normal range = 0.15 - 0.48 g/L). During the last year he had three episodes of angioedema treated with tranexamicacid.

The patient was treated with oral danazol 150 mg two times a day, 7 days before the surgical procedure and received a dose of C1-INH (Berinert ® P: CSL Behring GmbH, Marburg, Germany) 1,000 U iv 1 hour before surgery. After premedication with midazolam 3 mg and remifentanil 30 mcg iv, anesthesia was induced with propofol 180 mg and tracheal intubation was performed after providing cisatracurium 10 mg. Total intravenous anesthesia (TIVA) technique was performed with propofol 5 mg/kg/h and remifentanil 0.2-0.5 mcg/kg/min, with volume controlled ventilation, using bolus of cisatracurium 2 mg iv every 20 min. Intraoperative dexamethasone 20 mg i.v was provided. The patient underwent a L4-L5 posterior interbody arthrodesis with discectomy.

Anesthesia was uneventful during the whole procedure and he did not show any signs of anaphylactic reaction or edema. When the patient returned to the supine position, we found

some dermographism signs on the support areas (thorax and abdomen) of the spinal surgery support pad. Moved to the ICU where he remained for 24 hours stable without administering more C1-INH doses.

DISCUSSION

HAE is a rare disease characterized by self-limiting tissue swelling. These patients have recurrent episodes of edema mainly affecting the limbs, face, larynx and intestinal wall, causing intense pain, cramping, nausea, vomiting and diarrhea that may seem a surgical abdomen. The underlying cause of HAE is attributed to autosomal-dominant inheritance of mutations in C1-INH gene, which is mapped to chromosome 11 (11q12-q13.1), but the absence of family history does not rule out the possibility of HAE and 15% of cases are due to spontaneous gene mutation. C1-inhibitor protein is responsible for inactivating C1, which is the initiator of complement activation by the classical pathway. The absence of C1-INH leads to continuous activation C1, and C2 to C4 hydrolyzed which values are very low in these patients. The incidence is one in 10,000-50,000 people in the Unites States and the minimum prevalence in Spain is one in 100,000. Around 14% of these patients are asymptomatic [3]. Laryngeal edema is particularly serious and may endanger the patient's life. The episodes occur without an apparent cause, although anxiety, stress or minor trauma can trigger an attack. Our patient reported a tendency to coincide with cold episodes. The diversity of symptoms makes this disease difficult to recognize and it course could be varied and unpredictable. The long-term prophylactic treatment is usually done with danazol (analog of testosterone, increasing serum levels of C1 inhibitor and C4) and tranexamic acid (antifibrinolytic, probably its action on the C1 inhibitor activity depends on the antiplasmin action that regulates the release of vasoactive mediators) [4]. First line drugs for emergency treatment of laryngeal edema can be done with C1-INH as well as contact system modulators, such as ecallantide and icatibant. If first line drugs are not available, fresh frozen plasma (FFP) or solved detergent treated plasma (SDP) is the next choice.

A novel therapy are the Contact System Modulators, these drugs works as kinin pathway modulators. Two representatives of this class are Ecallantide (acts as a potent reversible inhibitor of plasma kallikrein) and Icatibant (which is a specific and selective competitive antagonist of bradykinin B2 receptor) [5,6].

The C1-INH preparation is available as both, human plasma derived and recombinant human protein (Rhucin in Noth America and in Europe, it is known as Ruconest: Pharming Technologies BV, Leiden, The Netherlands). The use of this preparation is indicated for the treatment of acute episodes of angioedema and as preoperative prophylaxis. In this last case, we should use a dose of C1-inhibitor 500-1000 U iv, 1 hour before surgery, and it is desirable that the patient has received hormonal therapy (danazol) in the week prior to surgery. In any case, we had available at least another 2 doses of C1-INH by hand, in case the patient has angioedema intra or postoperatively. After surgery, it is recommended these patients to transfer to the ICU for at least 24 hours for monitoring and early treatment in case of angioedema or other complication. In our case we chose general anesthesia technique in order to secure airway by tracheal intubation (TI) in case of laryngeal edema, considering that this procedure takes place necessarily in prone. Other authors chose regional anesthesia to avoid TI [7]. In spite of TI can be a stimulus capable of producing an episode of angioedema in these patients, we preferred to intubate him previously, since the patient kept all the intervention face down. We conclude that in patients with HAE, it is very important to adequate preoperative prophylaxis for edema acute attacks using C1-Inhibitor (1,000 U iv, 1 hour before surgery) and have some extra doses ready in case of need it perioperatively.

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