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Anesthesia for Urgent Surgery in a Patient with Angioneurotic Oedema: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Author SK designed the study, performed the case report writing, wrote the therapeutic protocol and wrote the first draft of the manuscript, author AB managed the literature searches, author HD established the plan of the article, author MM helped to achieve abdominal scan results and images in the operating room, author KAE coordinated the work, author SS corrected the article before submission. Author HB helped to translate from French to English and author AB supervised the work. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Hereditary angioneurotic oedema is an autosomal dominant disease associated with serum deficiency of functional C1-inhibitor. It is characterized by periodic swelling of subcutaneous tissues, abdominal viscera and upper airways. Lethal acute episodes of oedema can occur during anaesthesia and surgery. It is essential to prepare such patients before surgery. This article describes a case and the various preventive measures used to avoid acute episodes during anaesthesia for urgent surgery for mesenteric ischemia. In emergency situations where C1 inhibitor concentrate is not available, fresh frozen plasma (FFP) can be used as an alternative, as it also contains C1 inhibitor, corticosteroids, antihistamines, and epinephrine can be useful adjuncts but typically are not efficacious in aborting acute attacks. Prophylactic management involves long-term use of attenuated androgens or antifibrinolytic agents (Tranexamic acid).Their various indications are discussed.

Keywords: Angioneurotic oedema; c1-inhibitor deficiency; prevention; complication.

1. INTRODUCTION

Hereditary angioneurotic edema (HAE) is an autosomal dominant disease caused by a low level or improper function of a protein called the C1 inhibitor that afflicts 1 in 10000 to 1 in 150000 persons; HAE has been reported in all races, and no sex predominance has been found. It manifests as recurrent attacks of intense, massive, localized edema without concomitant pruritus, often resulting from one of several known triggers. However, attacks can occur in the absence of any identifiable initiating event. Hereditary deficits (autosomal dominant transmission) are owing to an abnormality in the C1 inhibitor gene resulting in a quantitative (type 1) or qualitative (type 2) deficit in C1 inhibitor. Acquired C1 inhibitor deficits are rare and are most frequently associated with lymphoproliferative syndrome. C1-Inh deficiency results in an increase in kallikrein, which in turn increases the production of bradykinin, an extremely labile mediator responsible for the onset of angioneurotic oedema [1]. Apart from the age of onset, most often in childhood in the hereditary forms and after 40 years in the acquired forms, the clinical presentation is similar regardless of the type [2]. The disease progresses by crises that can be triggered by stress, anxiety, even minimal physical trauma, surgery, The following observation illustrates what to do with a patient with this condition who needs to undergo an emergency operation due to Mesenteric ischemia. These therapies are intended to provide short-term prophylaxis, as well as treatment for the attack of a crisis of laryngeal or intestinal edema.

2. CASE REPORT

We report the case of a 35-year-old patient, who was suffering from angioneurotic edema diagnosed in childhood following a repetitive painful abdominal syndrome as well as peripheral subcutaneous edema with frequent but benign relapses, when her Laboratory testing revealed total C1-INH deficiency, known to have atopic eczema since childhood and acute systemic lupus erythematosus The last two pregnancies were conceived at term, and the deliveries were carried out in Spain by cesarean sections C1-INH and dexamethasone were administered preoperatively. She underwent tonsillectomy in childhood. No family history was found.

She presented to us with clinical picture of the surgical abdomen with bowel obstruction.

Clinical examination found a tender distended abdomen, a generalized defense, the bowel movement was absent, the hernial orifices were free, the rectal bulb was empty, Laboratory examinations revealed hyperleukocytosis (white blood count, 35000/uL) and increased C reactive protein (CRP) levels (463mg/L), liver function tests; ALAT 70 IU / L, ASAT38 IU / L, total bilirubin 53 mg / L, amylase 90 U / L, lipase 45 U / L, aerobic and anaerobic blood cultures were carried out, as well as a complementary infectious assessment. This biological examination thus objectified a biological inflammatory syndrome of infectious origin with functional renal failure. During the transfer to intensive care for monitoring, oxygen therapy, with peripheral venous catheter, right jugular central line, a right radial arterial line for blood pressure monitoring, and a urinary catheter were

taken, an empiric broad-spectrum antibiotic therapy based on ceftriaxone, Metronidazole, and Ciprofloxacin was started, rehydration, correction of hydroelectrolyte disturbances. Arterial gas analysis was performed showing pH at 7.25, PaCO2 at 29 mmHg, PaO2 at 150 mmHg, HCO3 at 18 mmol / L and a BE at -9 mmol / L, hyperlactatemia at 3.5 mmol / L. After effective volume expansion with relaunch of diuresis, the Computed tomography angiography (CTA) was performed showing peritoneal effusions suggesting acute generalized peritonitis and signs of digestive ischemia related to isolated thrombosis of the very superior mesenteric artery, (Fig. 1). Following these results, heparin therapy was started. The patient was admitted in the operating room, before the anesthetic induction, 3 units of PFC FFP were administered non-invasive monitoring was initially established, including non-invasive blood pressure (NBP), heart rate (HR), pulse oximetry (SpO2), and electrocardiography (ECG). Blood pressure monitoring. Urinary catheter was monitored. The initial vital signs were listed below: BP 90/40 mmHg; HR 114 bpm; SPO2 95%. Arterial blood gas showed: PH 7.30; PCO2 26 mmHg; PO2 160 mmHg; Na+ 134 mmol/L; K+ 3.8 mmol/L; Ca2+ 1.08 mmol/L; Glu 10.2 mmol/L; Lac 2.0 mmol/L; Hct 26%; BE -7.0 mmol/L; Hb 8.6 g/L. ECG showed sinus tachycardia of 114 bpm. Rapid induction of general anesthesia for intubation was conducted under epinephrine 0,5/ug/kg min to maintain a stable hemodynamic state, with midazolam 2 mg, fentanyl 0.2 mg, etomidate 10 mg, 1% propofol 60 mg, and Vecuronium 65 mg. After intubation, another shot of fentanyl 0.1 mg and midazolam 2 mg were given. Continuous infusion of 2% propofol 4-8 mg/kg/h and cisatracurium 10 mg/h intraoperative, with intermittent bolus of fentanyl were performed to maintain the depth of anesthesia and hemodynamic. The patient was steady during the surgery; arterial blood gas index was intermittent obtained to ensure the electrolyte balance and hemoglobin. The total amounts of intraoperative transfusion were LPRC 400 ml, plasma 200 ml, lactated ringer's solution 500 ml, succinylated gelatin 500 ml, and NaHCO3 solution 1 ml. At laparotomy cloudy and foul-smelling seroma effusion was found, the small intestine was necrotic, 25 cm after the duodenojejunal angle up to 35 cm in front of the ileocecal valve of Bauhin (Fig. 2), an extensive bowel resection with double stoma was performed leaving 60 cm of jejunum. The total urine output was 950 ml and blood loss was 200 ml. Subsequently she was transferred to surgical intensive care where heparin sodium anticoagulation was continued, extubated on the fifth day after a good evolution,

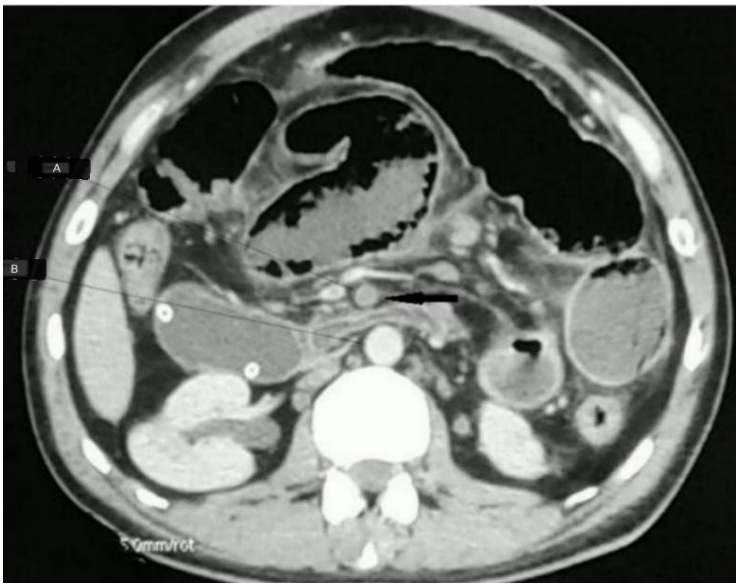

Fig. 1. Abdominal C + CT scan: very thin non-enhanced digestive wall indicating ischemia of arterial origin. Endoluminal defect not enhanced after injection: thrombus in the superior mesenteric artery (SMA) (arrow A) Free abdominal aorta (arrow B). Axial cut

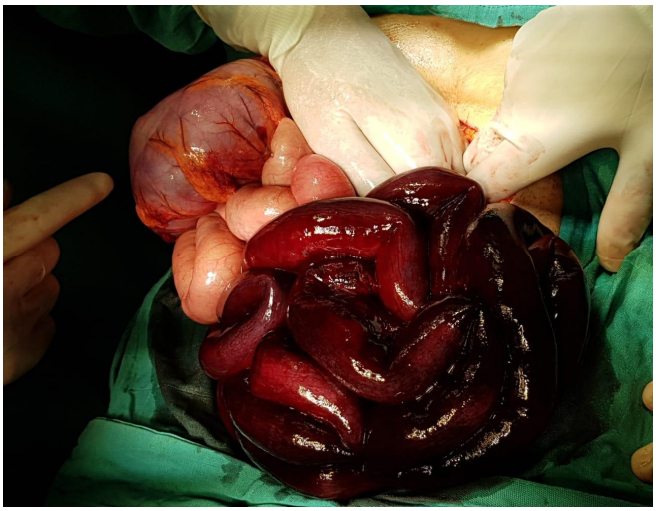

Fig. 2. Operative image showing extensive small intestinal necrosis in our patient

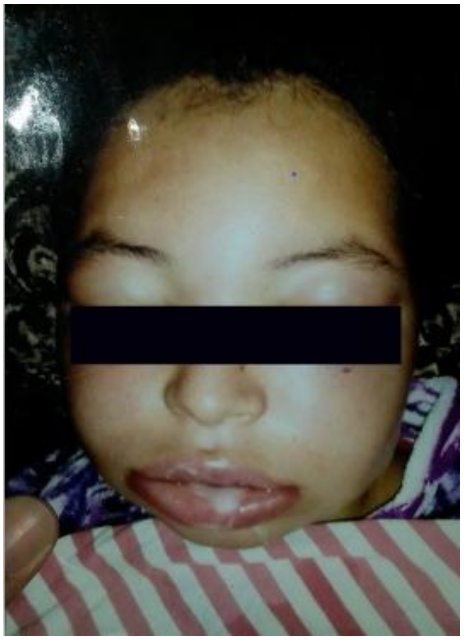

Fig. 3. Photo showing facial edema in our patient with C1 esterase deficiency, 30 min before her first reintubation

a few hours later, however, the patient presented the appearance of edema of the face and limbs associated with pharyngeal discomfort with dysphagia (Fig. 3). Despite the administration of inhaled corticosteroids, sudden inspiratory dyspnea developed, with prolongation of inspiratory time, indrawing and cornage, characteristic of high obstruction. On the other hand, we noted the absence of hemodynamic Reintubation quickly became necessary due to the ineffectiveness of the pharmacological treatments. This procedure was made difficult by the presence of a significant oropharyngeal edema visualized during direct laryngoscopy. In view of a suspicion of anaphylaxis, heparin treatment was withheld and anticoagulation was provided by administration of fondaparinux subcutaneously. Biologically, the dosage of C1 esterase inhibitor level was 80% (standards 70 - 130%), factor XII was also within normal values at 61% (standards 50–150%).The patient, still being treated with antibiotic therapy, was then

progressing quite favorably on the respiratory level and was extubated after 48 hours of mechanical ventilation. Unfortunately, a recurrence of high dyspnea on a new laryngeal edema was observed 30 hours after extubation, justifying a new orotracheal reintubation, the patient's subsequent evolution was fatal.

3. DISCUSSION

The clinical picture is marked by non-erythematous, non-pruritic and recurrent edema of the extremities, face, digestive mucous membranes (which may suggest acute abdominal syndrome), respiratory mucous membranes (which may progress to asphyxial edema of the glottis), [3]. The biological diagnosis is based on the demonstration of the antigenic deficit in quantitative C1INH (type 1): 85% of cases with a reduced blood level, less than 50% of the normal value) in most of the siblings. Some of them may have an antigenically intact but qualitatively non-functional protein (type 2): 15% of cases and, in this case, a functional assay is necessary to confirm the diagnosis [1,4]. This condition can pose serious problems for the anesthesiologist, since the crises are life-threatening and can be confused with anaphylactoid reactions or surgical abdominal emergencies hence the interest of long-term preventive treatment in order to reduce the frequency of those crises and improve the quality of life of these patients [5]. This treatment is based on specific therapies that are better and better codified (danazol, anabolic steroid, tranexamic acid, antifibrinolytic, and purified concentrates of C1 INH) [6,7]. Regular clinical and biological monitoring is always necessary to adapt the dosage of the preventive treatment.

For short-term prophylaxis, Patients should be seen at a distance from the procedure in preanesthesia consultation ; the preparation must be considered in collaboration with the internist who follows the patient and the notified blood transfusion center, in order to have a sufficient quantity of C1INH available for the perioperative period. If the patient receives long-term hormonal treatment, it should be continued until the day before the operation and resumed as soon as possible after the operation. Apart from dextrans [8], there are no drugs used in anesthesia that are formally contraindicated. Angiotensin converting enzyme inhibitors can trigger crises and should be excluded [8]. The intramuscular route should also be avoided for premedication [4]. The risk of anaphylactic manifestations is increased in patients with HAE. The anesthesia of the patient with HAE is a risky anesthesia, regardless of the procedure considered. Apart from absolute urgency, a preparation of 4 to 5 days, at best 8 to 10 days, with 600 mg / day of danazol, associated with an antifibrinolytic, is necessary if the level of C1INH remains low [8]. When preparation with danazol is not possible, or in an emergency, the infusion of C1INH concentrate, at a dose of 1,500 units in 20 min, must be carried out before the operation and possibly in the postoperative phase depending on the consequences, they have a therapeutic efficacy of 2 to 5 days [1,4,8]. When no preparation is possible and the C1INH concentrate cannot be readily available, at least 2 units of frozen fresh plasma (FFP) should be administered, which restores the level of C1INH for a period of 2 to 5 days [3] and 1 g of tranexamic acid before the operation,. Either way, danazol will be resumed as soon as the oral route become possible in the absence of contraindication. General and anesthesias are indicated [8]. The choice of technique should preferably be based on regional anesthesia techniques in order to avoid the trauma associated with intubation. All classes of hypnotics, opioids, curares and anesthetic gases can be used [9]. However, special care should be taken during intubation and oropharyngeal and tracheal aspirations [4,8] The use of the laryngeal mask has been reported. However, even a relatively large mask surface area can be a trigger for edema [9]. The urgency of the situation necessitated general anesthesia, and tracheal intubation was deemed essential. Despite its theoretical risks, FFP appears to be the mainstay of therapy and the transfusion was started as soon as possible, and an antihistamine was given. Fortunately, no edema developed postoperatively or during the first 5 days of resuscitation. Additional therapy might have been needed. The administration of tranexamic acid is not safe and haematological advice should be sought before use. All surgeries are possible, even those performed with cardiopulmonary bypass [9]. In this case, care should be taken with haemodilution which can lower the levels of C1 inhibitor and lead to complications. At the end of the surgery, the patients have to be monitored in a suitable environment, a crisis could still occur at a distance from the procedure. Outpatient surgery is contraindicated in this type of patient.

The risk of fatal laryngeal edema is maximum within 12 hours of the procedure. The

intervention can also trigger edema in the anatomical zone concerned by the act: urinary tract, uterus, skin ... In all cases, prophylaxis is necessary and the treatment of attacks must be available in the operating room and in post-intervention monitoring room [9]. Neither antifibrinolytic nor hormonal agents have shown a great benefit during acute attacks while FFP provide improvement within 40 minutes during acute attacks of ANO HAE [10]. The response to epinephrine, steroids and antihistamines is uncertain, although during life-threatening attacks these agents are often used [10]. Indeed, it has been suggested that high doses of adrenaline (1 mg subcutaneously per hour for a total of 11 mg) may be effective when FFP are not available [10]. Such therapy resulted in subjective and objective improvement within 6 hours, as was the case with our patient after her first attack of edema. The use of such high doses of catecholamine in the presence of airway obstruction with hypoxia and hypercapnia could be dangerous compared to the use of FFP which have been shown to be more effective [10]. It is essential to secure the upper airway in the event of obstructive edema, preferably by tracheal intubation, if not at worst by tracheostomy. If possible, laryngeal edema should be assessed, using indirect laryngoscopy when possible. Partial airway obstruction is quickly managed by initiating specific ANO HAE therapy and traditional edema therapy (epinephrine, etc." which should be discussed. Edema of the intestinal wall in acute attacks of ANO HAE can cause abdominal pain, nausea, vomiting, toxic megacolon and sometimes unnecessary laparotomies [10].

4. CONCLUSION

The originality of this observation lies in the fact that it was not possible either to carry out the classic preparation with danazol or with tranexamic acid or to infuse C1INH concentrates which was unavailable, only the infusion of FFP made it possible to perform this procedure without immediate complications. In patients with ANO, locoregional or general anesthesia can therefore be performed with a very low risk of complications, if simple prophylaxis is performed. At best, a preparation is needed several days before the planned intervention using danazol, in case this preparation is not possible a rapid preoperative infusion of C1INH is sufficient when they are available, otherwise an infusion of FFP is an effective therapeutic alternative in emergency situations. Thanks to these products, to close monitoring and a good knowledge of the pathophysiology of this condition, acute crises can be avoided during the perioperative period.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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