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Case Report
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Anaesthetic Approach for Patient with Hereditary Angioedema

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Abstract

Hereditary Angioedema (HEA), a disease caused by a mutation in the gene that encodes for the production of the fraction C1 in the complement (C1-INH), is a rare pathology (1/50.000) that causes swelling of the skin and submucosa in various organs, either naturally triggered or provoked by physical and psychological traumas, infections, or by the use of nonsteroidal anti-inflammatory drugs (NSAIDs) and angiotensin-converting enzyme inhibitors (ACEIs). Surgical trauma may spur the HEA crisis, leading to complications such as the swelling of the respiratory tracts and hemodynamic instability. Thus, the pre-surgical approach to HEA patients requires a specific plan that ensures short term prophylaxis, careful intra-operative management, rescue therapy and intensive post-surgery care. We present a report on a video-laparoscopic cholecystectomy approach for a 28-year-old woman diagnosed with asthma and HEA with symptomatic choledocholithiasis. We opted for short-term prophylaxis and immunology with the intravenous application of C1-INH. Ultrasonography imaging showed arterial wall oedema, which could correspond to a manifestation of C1-INH deficiency in the wall of the manipulated arteries during ultrasonography-guided puncture. Once the patient recovered consciousness, she was transferred to the intensive care unit and was discharged on the 6th day of hospitalisation.

Keywords: Anaesthesia, C1 complement inhibitory protein, hereditary angioedema

Introduction

Hereditary angioedema (HEA) is a dominant-autosomal transmitted, recurrent, and rare disease (1/50.000-100.000) caused by a mutation in the gene that encodes for the production of the C1 fraction inhibitor of complement (C1-INH) (1-4). This implies the reduction of classical and lectins tracts in the complement system and other proteases, clotting factors (XII and XI), and plasmin. The deficiency of C1-INH results in an over-activation of the contact system (Kinin-Kallikrein system), increasing the production of bradykinin.

The main clinical symptoms of HEA involve skin and submucosal swellings in various organs (2, 5). A crisis may arise naturally or be triggered by physical or psychological traumas, infections, or by the use nonsteroidal anti-inflammatory drugs (NSAIDs) and angiotensin-converting enzyme inhibitors (ACEIs). Perioperative care of HEA patients requires a specific plan that ensures short-term prophylaxis, careful intra-operative management, rescue therapy and intensive post-surgery care. The aim of this work is to describe the anaesthetic approach in a HEA patient in need of surgery.

Case Presentation

Informed consent was obtained from the patient before recording the information in this report. A 28-year-old woman (weight: 60 kg, height: 1.63 cm) was diagnosed with asthma and HEA type I and was proposed for video-laparoscopic cholecystectomy. In the pre-anaesthetic investigation, a positive family history was identified. The patient had had recurrent hospitalisations over the previous 20 years due to this crisis. She was tracheostomised after having developed tracheomalacia on account of long periods of orotracheal intubation. During the most severe episodes, she received fresh plasma for treatment and presented with transfusion reactions including fever, seizures

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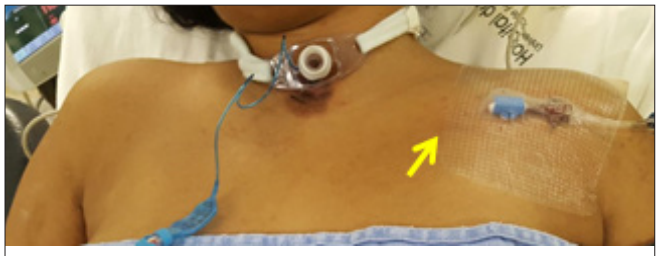

Figure 1. Infra-clavicular oedema

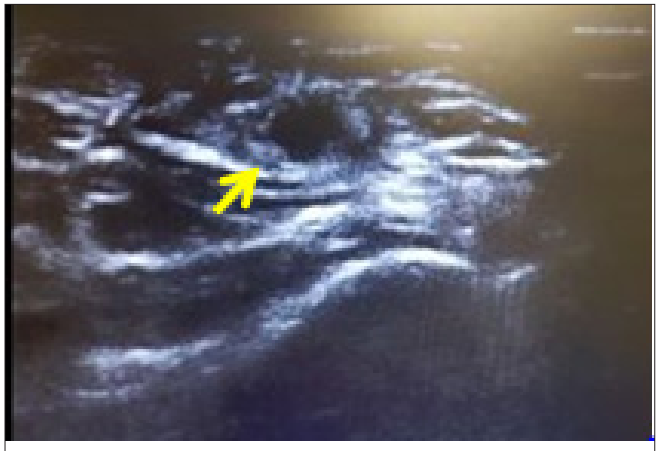

Figure 2. Transverse section of the radial artery showing increased arterial wall thickness (oedema)

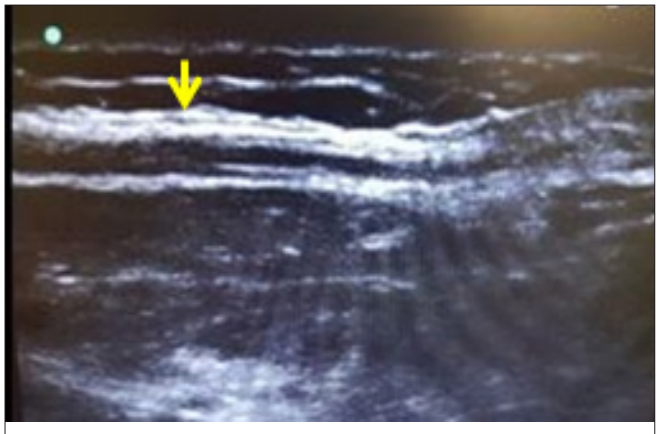

Figure 3. Longitudinal section of the radial artery showing increased arterial wall thickness (oedema)

and anaphylaxis. After discussing her case, we opted for short-term prophylaxis and immunology in the form of intravenous application of C1-INH.

On the day of the surgery, central venous access puncture was performed in the left sub-clavian vein, while observing infra-clavicular oedema of the manipulated region from the beginning of the puncture (Figure 1). Following intravenous administration of 1500 U of C1-INH concentrate 1 hour before the procedure, anaesthesia was induced with Fentanyl 4 mcg kg^{-1}, Sevoflurane 1.2 CAM, and Atracurium 0.5 mg kg^{-1}. After anaesthetic induction, attempts were made to puncture the radial artery, but there were cannulation difficulties. Ultrasonography-guided puncture was used when the thickness of the radial artery wall was increased. Imaging showed arterial wall oedema, which corresponded to a manifestation of C1-INH deficiency in the wall of the manipulated arteries (Figures 2 and 3).

Anaesthesia was maintained with 1 MAC sevoflurane and atracurium according to neuromuscular monitoring. The patient remained stable during the procedure, observing light loop oedema during video-laparoscopy. Once the patient recovered consciousness, she was transferred to the intensive care unit. During the first hours of ICU stay, she presented with dyspnoea and hoarseness but showed a good response to treatment with 30 mg of Icatibant that was applied subcutaneously. She was discharged on the 6th day of hospitalisation.

Discussion

Hereditary Angioedema is an autosomal dominant disease, which results from a biochemical abnormality in the levels or functionality of C1INH (4), a serial protease that blocks the activity of several proteases of the complement system (C1r, C1s, MASP 1 and 2), contact system (Kallikrein, factor XII), clotting (factor XI, trombina), and the fibrinolytic system (plasminogen activator, plasmin) (6). There is evidence of activation of several systems during HEA attacks such as the bradykinin-kallikrein system, factor XII fibrinolytic cascade and the dependent and complement systems. C1-INH deficiency results in excessive consumption of C4 and C2, in addition to excess production of bradykinin that binds to its B2 receptor (BDKRB2) from endothelial cells, increasing vascular permeability and inducing angioedema. Bradykinin also stimulates the production of nitric oxide, which triggers vasodilation by contraction of the cytoskeleton.

The diagnosis of HEA should be suspected when there is history of recurrent non-pruritic angioedema (7, 8). In our case report, a positive family history was identified and recurrent hospitalisations over the previous 20 years due to crisis since adolescence were noted. In terms of laboratory tests, serum levels of C4 can be used for screening. In addition, a quantitative and/or functional evaluation of C1-INH should be performed (2, 8).

Recommended strategies for the HEA therapy include long-term prophylaxis, short-term prophylaxis to patients who are likely to initiate an attack, and active treatment of the angioedema attacks to all patients. Long-term prophylaxis can be done with antifibrinolytics (epsilonaminocaproic acid and tranexamic acid), androgens (danazol, estazolol, and oxandrolone), plasma-derived C1-INH concentrate (PdC1-INH), and Cinryze and Berinert.

For short-term prophylaxis, the risk associated with the procedure and the availability of treatment for the acute HEA crisis should be considered. In higher-risk procedures or requiring intubation, PdC1-INH should be applied 1-6 hours prior to intubation. Fresh frozen plasma containing C1-INH is another pre-surgical option (8, 9). In this case, we opted for prophylaxis with intravenous application of C1-INH. On the day of surgery, 1 hour before the procedure, the patient received intravenous administration of 1500 U of C1-INH concentrate, which did not prevent the formation of oedema in the walls of the punctured radial artery during invasive blood pressure monitoring.

There are three medications available for acute crisis therapy: PdC1-INH, icatibant (bradykinin receptor blocker), and ecallantide (kallikrein inhibitor). Icatibant was used in our case to treat an acute crisis in ICU during the first few postoperative hours, which delivered an excellent response to the treatment. PFC can be used for emergency treatment. However, a paradoxical worsening may occur in some cases due to increased bradykinin.

If possible, the anaesthesiologist should prefer locoregional anaesthesia techniques to avoid manipulation of the respiratory tract (1, 3). In the case described, the patient had been previously tracheostomised, general anaesthesia was required, and short-term prophylaxis with Berinert was performed 1 hour before the incision. As currently recommended by HEA consensus, PdC1-INH has been used for prophylaxis of HEA crisis (9). With the recent availability of new therapies specific for HEA, it is expected that the incidence of acute exacerbations followed by perioperative complications will decrease.

Conclusion

Hereditary angioedema is a rare disease with serious complications that can lead to tragic situations during anaesthetic-surgical procedures. The anaesthesiologist has a primary role in the management of the patient's respiratory tracts and should value safety, opting for the most appropriate prophylaxis for each case.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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