

"Balancing Safety and Stress: Anesthetic Strategy in Pediatric Hereditary Angioedema"

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Introduction

Hereditary angioedema (HAE) is a rare disorder of C1-esterase inhibitor quantitative (type 1) or qualitative (type 2) deficiencies; characterized by episodic, non-histaminergic angioedema, which can involve the GI tract and genitals (may be misdiagnosed as a surgical abdomen), extremities and upper airway. Triggers include physical or psychologic stress and trauma. General anesthesia poses significant risk in HAE from airway compromise. Neuraxial anesthesia is a safe alternative but has technical and sedation-related challenges in pediatrics. This case highlights anesthetic management balancing safety and stress in a pediatric patient with HAE undergoing emergent surgery.

Purpose

Recognize the anesthetic risks in pediatric patients with hereditary angioedema and apply evidence-based strategies for anesthetic management, pharmacologic prophylaxis, and intraoperative vigilance.

Case Description

12-year-old boy HAE (treated with lanadelumab, a kallikrein inhibitor) and IDDM presenting with testicular torsion. We sedated with propofol (250 mcg/kg/min) then placed 1 mL of i.t. 0.75% h. bupivacaine; acetaminophen, ketorolac for postop analgesia. Uneventful orchiectomy, inguinal exploration, hernia repair, and fixation of contralateral testis. Prn lanadelumab was not needed.



Figure 1. Facial and extremity swelling in hereditary angioedema

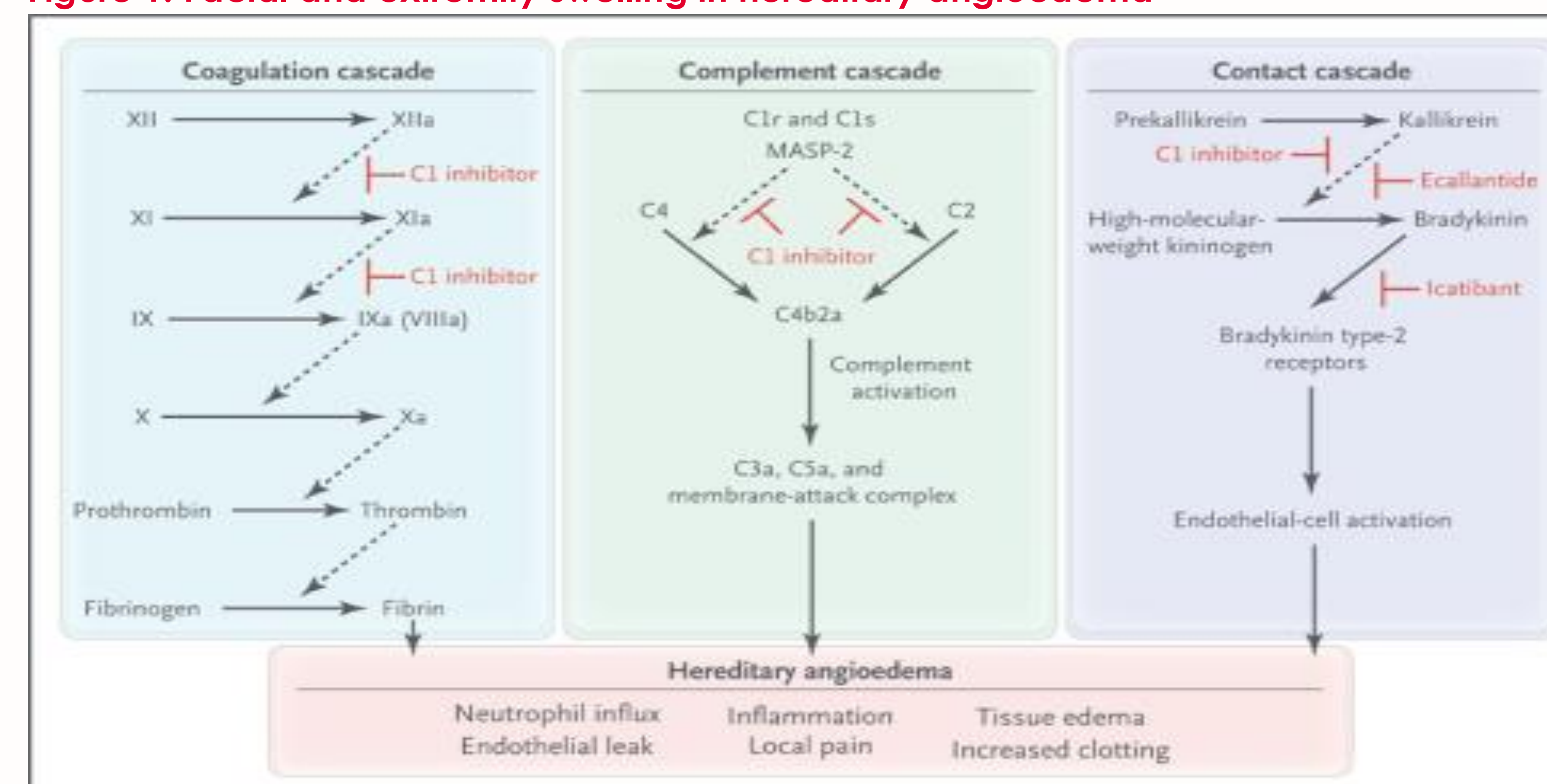


Figure 2. Dysregulation of Coagulation, Complement and Contact cascades in hereditary angioedema

Discussion

Treatments for anaphylaxis will not be effective in HAE, but HAE responds to prophylactic kallikrein inhibition (e.g. berotralstat, lanadelumab) as well as for acute treatment. Spinal anesthesia with sedation is an alternative to GA in pediatric HAE, balancing cooperation with preservation of the airway, and may include post-spinal epidural for extended procedures: This necessitates good communication with the surgeon to outline concerns and expectations.

Key points

1. HAE treatment is different to that of anaphylaxis
2. Regional anesthesia with sedation is a safe anesthetic approach in pediatric patients with HAE.
3. Careful perioperative planning and readiness to accommodate changes to plan are essential

References

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3. Image source: "healthjade.net/hereditary-angioedema"
4. Image source: "Morgan BP. Hereditary angioedema--therapies old and new. N Engl J Med. 2010 Aug 5;363(6):581-3.