



Introduction

Hereditary angioedema (HA) is a rare autosomal dominant condition with an estimated prevalence of 1/50,000 to 1/100,000 individuals¹. The pathophysiology of the disease results from decreased levels and/or function of C1 esterase inhibitor leading to unregulated activity of the complement cascade⁴. This activity results in episodes of subcutaneous and submucosal edema which can lead to life-threatening upper airway obstruction.¹ HA episodes can occur spontaneously or be triggered by pain, stress, trauma, and estrogen.² Given the association of these triggers with pregnancy it is unsurprising that these women will experience increased frequency of attacks during pregnancy³.

Case Report

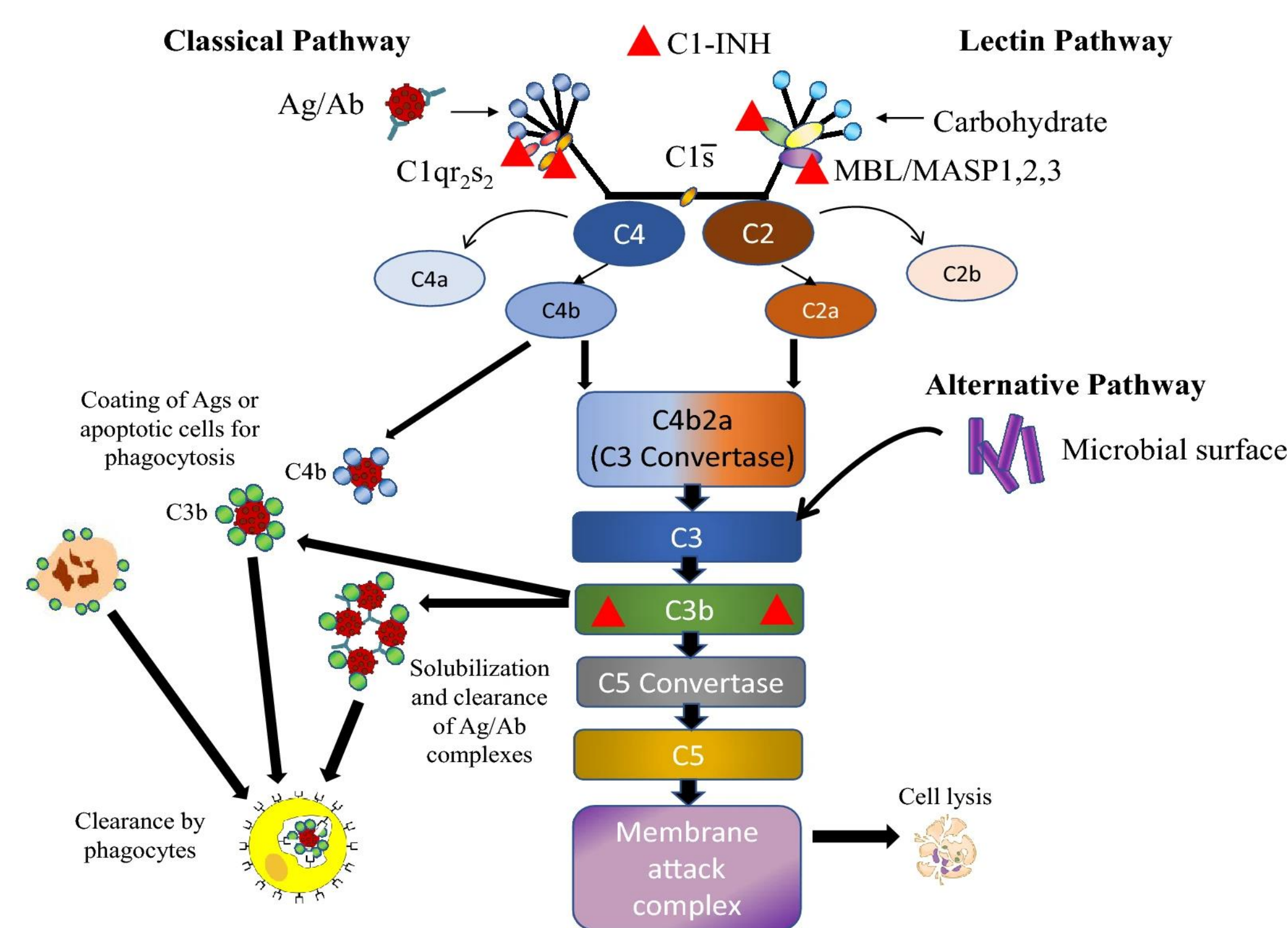
30 y/o G2P1011 female at 39 WGA who presented for induction of labor. The patient's medical history was significant for HA. The patient was diagnosed at the age of two and her attacks were associated with hormonal changes. During pregnancy she had increasing episodes with increased severity, many of which included laryngeal edema. These were thought to be triggered by increased estrogen levels, so she was started on a pooled human plasma C1 esterase inhibitor, for prophylaxis. She was seen in our perioperative clinic to determine best management for labor. She was advised to take her C1 esterase inhibitor on the morning of induction, to bring her rescue medications (bradykinin inhibitor and recombinant C1 esterase inhibitor) with her, and to consider an early epidural. On the morning of her induction, anesthesia was notified of her arrival, she received a prophylactic dose of her pooled human plasma C1 esterase inhibitor, her rescue medications were placed at bedside, a fully stocked emergency airway cart was placed outside of her room, and an early CSE was placed to prevent pain that could potentially trigger an episode. Labor was uneventful with a vaginal delivery of a healthy neonate.

Discussion

HA results from a deficiency of the C1 esterase inhibitor protein, causing unregulated activity of the complement cascade and overproduction of bradykinin that leads to edematous episodes that characterize the disease.¹ Treatments for the disease range from long- and short-term prophylaxis to rescue treatments for acute flares. The literature describes several cases of HA in pregnancy including two cases of uncomplicated vaginal deliveries where one received prophylactic medication and a case of uncomplicated cesarean delivery.^{2,3} There is also a case series of 125 deliveries, where 14 received short-term prophylaxis at the time of delivery, however only 13 received prophylactic treatment prior to pregnancy.⁴

While there are no formal guidelines on management in parturients with HA, prior evaluation and preparation by anesthesia providers is crucial, in addition to multidisciplinary delivery planning to minimize stressors. Early neuraxial analgesia should be advised, rescue treatments should be immediately available in the labor and operating rooms, and emergency airway equipment easily accessible. While the literature describes many cases of women successfully delivering without prophylactic treatment, an angioedema episode can have severe consequences in parturients whose airways may already be altered due to normal changes of pregnancy. Immediate availability of C1 inhibitor concentrates, bradykinin antagonists and kallikrein inhibitors could be lifesaving in case of an emergency.

Figure 1: The Complement Cascade



Adopted from: Levy, D, et al. (2020). *Allergy, Asthma & Clinical Immunology*, 16(1), 1-8.

References

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