ACQUIRED HAEMOPHILIA A PRESENTING AS ACUTE AIRWAY OBSTRUCTION

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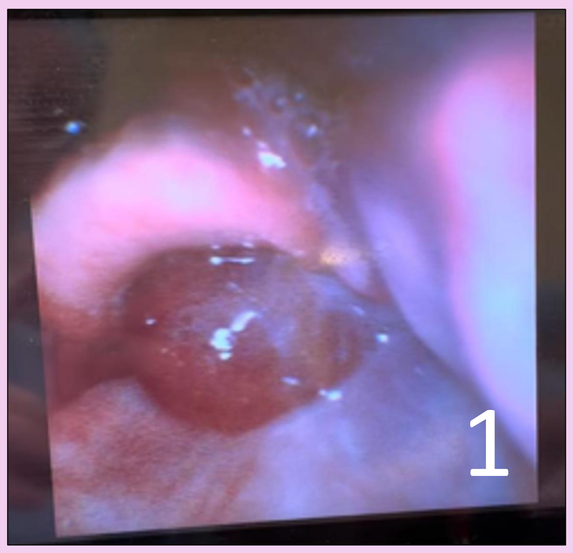
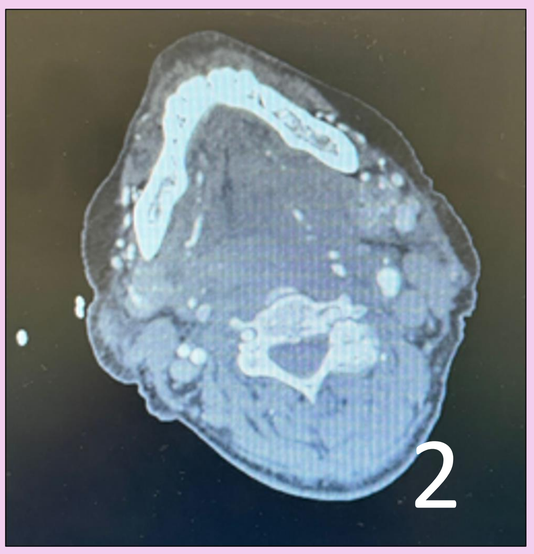
Introduction

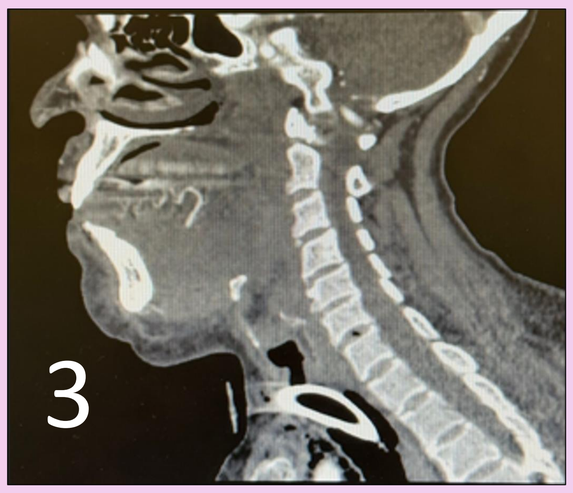
Acquired haemophilia A (AHA) also known as factor 8 deficiency is a rare autoimmune bleeding disorder in which antibodies to factor 8 develop in a person with previously normal haemostasis. (1) (2) This case discusses an 88 year old female presenting with rapidly progressing ecchymosis to the neck and significant swelling under the tongue requiring emergency tracheostomy under local anaesthetic due to impending airway obstruction caused by AHA.





Case Report Information

- 88 year old female patient
- PMH: mixed dementia, hypertension, lymphoedema, previous right MCA infarct, hypothyroidism
- DH: furosemide, levothyroxine, clopidogrel, atorvastatin, donepezil, ramipril, lansoprazole
- Brought to emergency department (ED) by family due to dysphonia and reported dysphagia. Stable on arrival but moved to resus due to rapidly progressive airway occlusion.
- Flexible nasoendoscopy (FNE) showed a large haematoma partially obstructing the airway causing clinical deterioration and patient becoming unable to swallow saliva.



Images

1-Direct video laryngoscopy post tracheostomy
2&3-Computed tomography scan post tracheostomy

Interventions

- Category 1 transfer to theatre for emergency tracheostomy under local anaesthetic Performed under decision of best interest in collaboration with family
- Formal panendoscopy post tracheostomy showed significant pharyngeal haematoma and an airway that would not have been successfully intubated
- Computed tomography scan post theatre – large left sided sublingual, submental, submandibular and pharyngeal haematoma with no identifiable bleeding point
- Treatment with 11 days of Obizur (porcine factor VIII) 6000U and total of 1 month of corticosteroids initially methylprednisolone 40mg
- Successful decannulation ten days post presentation
- Following decannulation started on mycophenolate mofetil for immunosuppression
- Completed four weeks of rituximab
- Discharged home 32 days after initial presentation

Discussion

Early intervention and close monitoring were essential in ensuring a positive outcome. Due to the patient's dementia there was a poor history therefore, the prompt recognition in ED of the potential for airway compromise despite no clear diagnosis allowed careful monitoring and early recognition of deterioration. The rapid decline and increase in severity of symptoms over a short time frame dictated the decision to take to theatre prior to discussion with haematology. However, it would be recommended to involve haematology immediately to allow for intervention prior to surgery and ensure treatments are initiated promptly. (3) Whilst acknowledging the risk of taking to theatre prior to haematology input steps were taken to minimise bleeding whilst operating notably the use of monopolar over cold steel dissection.

Literature

At time of this case occurring there were 16 documented cases of AHA presenting as acute airway obstruction requiring emergency tracheostomy. Of these only one patient did not survive their admission. All patients required a definitive airway with most requiring a tracheostomy prior to medical treatment that included factor VIII and steroids.

Conclusion

Airway obstruction secondary to AHA can be fatal. Early recognition of symptoms, awareness of its rapidly progressive nature and immediate haematology input is paramount to improve outcomes. Good surgical technique is required to minimise bleeding and it is recommended that the most experienced surgeon available performs the operation.(4) Symptoms patients may present with in AHA may be initially subtle and awareness of these allows appropriate speciality referrals and timely intervention.

Acknowledgements

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References: (1) NORD (2026). Acquired Hemophilia A [Online]. Available at https://rarediseases.org/rare-diseases/acuired-haemophilia/ [Accessed 12 April 2026] (2) Berger, K. and R. Parker (2019). "Acute respiratory failure and carotid baroreceptor-mediated bradycardia due to spontaneous neck hematoma in acquired hemophilia A." American Journal of Respiratory and Critical Care Medicine 199(9). (3) Srivastava, A. et al. (2020). "WFH Guidelines for the Management of Hemophilia, 3rd edition." Haemophilia. 26(Suppl 6): 1-158. (4) Tiede, A. et al. (2019) "Clinical evaluation of bleeds and response to haemostatic treatment in patients with acquired haemophilia: A global expert consensus statement." Haemophilia. 25:969–978.