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**INTRODUCTION**

- Acute compartment syndrome (ACS) is a surgical emergency and usually occurs following trauma.
- Non-traumatic causes are less common and are due to ischemia-reperfusion injury, bleeding disorders, nephrotic syndrome, animal bites, and rarely hereditary angioedema.

**THE CASE**

24 year old African American female presented with left hand swelling and darkish discoloration over the past 12 hours.

**PMH**

- DVT and pulmonary embolism with prothrombin 20210 mutation
- Hereditary angioedema, type 3 (over 20 exacerbations/complications)

**Medications**

- Loratadine
- Ranitidine
- Risperidone

**Family History**

- Hereditary angioedema; CAD

**Physical Examination**

- 4SS except for tachycardia
- General: Pleasant, cooperative, in mild distress
- HEENT: no facial or neck edema; periorbital scar and multiple facial scars
- CV: palpable pulse in radial and ulnar position on left hand
- MSK: Left upper extremity markedly edematous with marked tenderness on palpation from left hand to mid forearm; marked discoloration of her left thumb - dark purple and well demarcated.
- Skin: tense mostly in the left distal arm
- Neuro: decrease in sensation in subcutaneous tissue overlying left thumb

**FURTHER INVESTIGATIONS**

**Left Upper Extremity Arterial Duplex**: negative for thrombosis or hemodynamic stenosis

**Ultrasound of LUE (non-vascular)**: Diffuse subcutaneous and muscle edema without drainable fluid collection. Findings may represent reactive edema, ecchymosis or cellulitis

**LUE Venous Ultrasound**: Negative for DVT

**Left Wrist X-ray**: Negative for fracture

**Erythrocyte Sedimentation Rate**: 65

**NEXT STEPS**

Treatment with C1 esterase-inhibitor, steroids, H2 blockers was not effective and 1 day after initial change, there was mottling of thumb and thenar skin

Urgent Plastics/Hand Consult: Thenar compartment had 42 cm of water pressure → Urgent Fasciotomy

Fasciotomy: Mottling of ulnar aspect of left thumb, small artery was clotted in proximal aspect of left thumb

**DISCUSSION**

The final diagnosis was acute compartment syndrome due to underlying HAE or untreated thrombotic disorder

Hereditary angioedema, type 3: Rare form which does not have deficiency of C1-INH. Usually normal C3/C4 and seen in females. Exacerbations usually flared by pregnancy or oral contraceptives containing estrogen.

**REFERENCES**

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**CONCLUSION**

- Second reported case of compartment syndrome of hand caused HAE, type 3
- Patients with HAE most commonly present with angioedema flares, but this case demonstrates that closely reassessing the patient either in the emergency room or on the medical floor and performing necessary diagnostic interventions can help prevent loss of a distal extremity.

**TREATMENT AND PROGNOSIS**

- Urgent fasciotomy
- Multiple debridements to salvage thumb
- Creation of the bipedicle flap
- Heparin (or other anticoagulant) for prevention/treatment of thrombotic disorder
- Antibiotics based on cultures
- Hand therapy
- Even with treatment, prognosis to regain full function is low because >2 days to alleviate pressure (only 75% function after 6 months)

**DISCLOSURES**

- There are no financial conflicts for any of the authors.