# Acquired Hemophilia A following COVID-19 infection

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## BACKGROUND

- Acquired Hemophilia A (AHA) is a rare condition characterized by development of inhibitory antibodies to Factor VIII.
- Idiopathic in most cases, however, could be associated with malignancies, infections, drugs, autoimmune conditions.
- In recent years, cases of AHA following COVID-19 vaccination have been reported and AHA following COVID-19 infection is thought to be rare.

## CASE REPORT

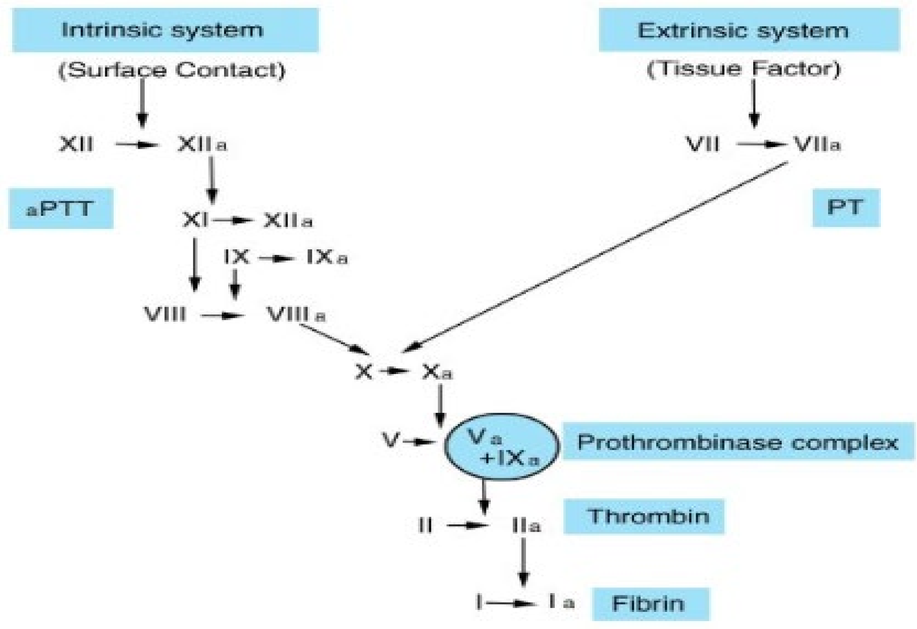
- A 69-year-old gentleman with history of congestive heart failure and dilated cardiomyopathy presented to an outside facility with exertional dyspnea.
- Hb was 5.8 g/dL (baseline Hb 13-15 g/dL). COVID-19 PCR was positive on admission, but he had no respiratory symptoms.
- On admission he was noted to have spontaneous intramuscular hematoma over the right chest wall, right upper extremity, and right masseter muscle. He required 8 units of PRBC transfusion over next few days.
- 10 days later, PTT was elevated at 140 s (Ref range 27.6 - 37s); PT/INR was normal.
- He was transferred to our facility for a tertiary level of care.

## DIAGNOSIS

| Investigation | Value | Reference |
|---|---|---|
| PT | Normal | |
| PTT | 140s | 27.6-37s |
| PTT mix 1:1 | 70s | 25-37s |
| Factor II | 74% | 50-150% |
| Factor V | 91% | 50-150% |
| Factor VII | 103% | 50-150% |
| **Factor VIII** | **<1%** | **50-150%** |
| Factor IX | 20% | 50-150% |
| Factor X | 59% | 50-150% |
| Factor XI | 29.8% | 50-150% |
| Factor XII | 3% | 50-150% |

Factor VIII activity assay <1%; Factor VIII inhibitor level was 677 Bethesda units (BU) confirming AHA. Activity assay for factors IX, XI, and XII were normal.

## COAGULATION CASCADE




## MANAGEMENT

- Initiated on prednisone 1mg/kg/day, Factor Eight Inhibitor Bypass Activity 100 units/kg every 12 hours for 4 doses. Cyclophosphamide initiated at 50mg/day and increased to 100mg/day the following day. There was no extension of hematoma and hemoglobin remained stable. He was discharged on prednisone and cyclophosphamide.
- 2 weeks later, during an outpatient visit, Factor VIII inhibitors had down trended to 288 BU, showing inadequate response to cyclophosphamide. Weekly Rituximab 375mg/sq meter was added to his treatment regimen.
- 2 weeks later, he suffered a cardiac arrest with successful resuscitation and admission to the critical care unit for management of septic shock and hemorrhagic shock. Hospitalization complicated by C Diff colitis, CMV viremia, and neutropenia.
- Though initially held, Rituximab and Cyclophosphamide were resumed due to active bleeding and worsening hematomas. Following a complicated hospitalization course, he succumbed to multi-organ failure.

## CONCLUSION

- Immune dysregulation following COVID-19 infection has been suggested as a pathophysiology in the development of AHA. In these patients, Naranjo score of probability is 5.
- In our review of the literature, less than 10 cases of AHA have been reported following COVID-19 infection. Recent literature suggests this diagnosis is often delayed by not checking coagulation tests as happened in our patient.
- Our patient serves to highlight that there is no direct correlation between the severity of COVID-19 infection and severity of AHA and to raise awareness of this rare entity following COVID-19 infection.

## REFERENCES