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Thrombotic Thrombocytopenic Purpura Presenting as an Acute Coronary Syndrome

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Introduction

- Thrombotic thrombocytopenic purpura is a thrombotic microangiopathy caused by severely reduced activity of the von Willebrand factor-cleaving protease ADAMTS13. It is considered as a medical emergency and could be fatal if appropriate medical management if not initiated in time. Rarely, it can present as acute coronary syndrome.

Case Presentation

- We present a case of an 80-year-old female with a past medical history of hypertension and irritable bowel syndrome who presented to the emergency department with chest pain. Her chest pain was centrally located, exertional and relieved by sublingual nitroglycerin. The patient also reported having dizziness, generalized fatigue and headache. On arrival, she was noted to be hypertensive (158/82) but otherwise hemodynamically stable. Physical examination was unremarkable. Laboratory evaluation revealed elevated high sensitivity troponin with an initial value of 434 ng/L (reference range <19 ng/L) and peaking at 817 ng/L, elevated creatinine of 1.61 mg/dL (reference range <1.16 mg/dL), normocytic anemia of 11.2 g/dL (reference range 12.0 - 15.0 g/dL) and thrombocytopenia with a platelet count of 53 K/uL (150 - 450 K/uL). An EKG was obtained showing T-wave inversion in V4, V5 and V6 with poor R-wave progression from V1 to V3 (figure 1). She was admitted for further management of a presumed non ST segment elevation myocardial infarction (NSTEMI) and was given heparin and aspirin. Two days after admission, her platelet count and hemoglobin dropped to 19 K/uL and 10.2 g/dL, respectively. Further laboratory work up revealed a decreased haptoglobin less than 30 mg/dL (reference range 30 - 200 mg/dL), elevated lactate dehydrogenase (LDH) of 484 IU/L (reference range <250 IU/L), elevated total bilirubin of 1.9 mg/dL (reference range <1.2 mg/dL) with normal direct bilirubin of 0.2 mg/dL (reference range 0 - 0.3 mg/dL) and a negative direct coombs test. A peripheral smear was obtained showing mild schistocytosis. An ADAMTS13 level was obtained showing decreased values of <5. The patient's heparin was discontinued and she was started on prednisone and plasmapheresis with improvement of her platelet count. Her clinical symptoms improved as well and she was started on rituximab for relapse prevention.

Discussion

- Thrombotic thrombocytopenic purpura (TTP) is described by the classic pentad of microangiopathic hemolytic anemia, thrombocytopenia, renal involvement, central nervous system manifestations and fever. Our patient presented with chest pain and neurological symptoms and was found to have anemia, thrombocytopenia and an elevated creatinine. Those findings with an elevated troponin and EKG changes should raise the suspicion of TTP-related acute coronary syndrome. The exact mechanism of how TTP can cause acute coronary syndrome is not well understood, especially that this has been rarely described but is thought to be secondary to thrombosis of the small vessels supplying the myocardium.
- Given its rarity, there is no consensus regarding the exact management strategy but emergent plasmapheresis should be started to improve blood counts and improve clinical symptoms. Coronary angiopathy should be deferred until blood counts improve and as it may exacerbation TTP. For our patient, immediate initiation of plasmapheresis and steroids corrected her laboratory abnormalities and relieved her symptoms.

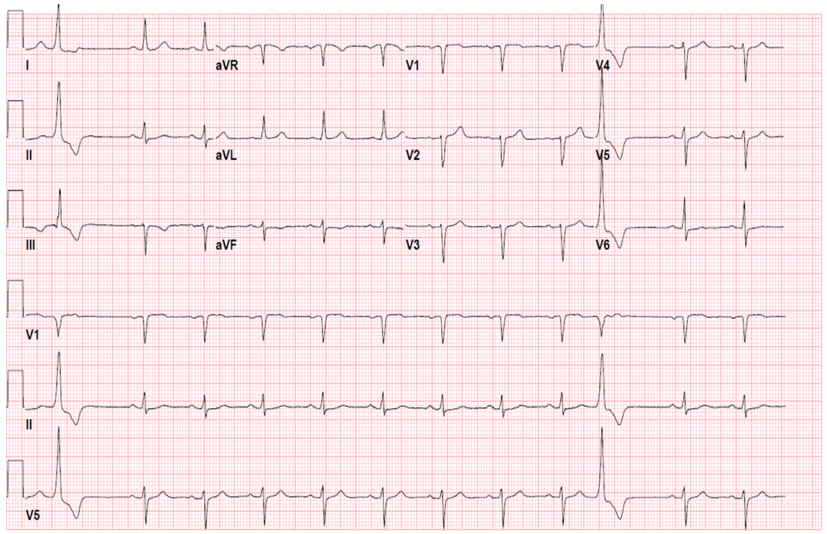

Figure 1. Initial electrocardiogram our patient presented with.

Conclusion

- This case represents a rare disease process that required plasmapheresis and immunosuppressives.

References

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