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Background

- %Thrombotic thrombocytopenic purpura (TTP) is a thrombotic microangiopathy caused by decreased activity of ADAMTS13, a protease that cleaves von Willebrand factor. Acquired TTP is due to an autoantibody against ADAMTS13.
- %Pentad: fevers, altered mental status, thrombocytopenia, reduced kidney function, and MAHA[1]. Neurologic, cardiac, and/or renal abnormalities may or may not be seen as well[2].
- %Cardiac involvement may present as hypertension, myocardial infarction, both ST elevation myocardial infarction (STEMI) and non-ST elevation myocardial infarction (NSTEMI)[3,4], atrial fibrillation, or congestive heart failure[1,5].
- %The most common cause of death associated with TTP is due to cardiac arrest or myocardial infarction, thought to be due to microthrombi[6,1].
- %Plasma exchange removes the anti-ADAMTS13 antibodies[1]. Other therapies include glucocorticoids, rituximab, and caplacizumab in certain high-risk patients[1].

Case Presentation

HISTORY OF PRESENTING ILLNESS:

A 64-year-old African American male presented complaining of new onset, “sharp”, left sided chest pain. It initially occurred in the evening while he was walking in his back yard, but improved with rest. However, another episode occurred about five hours later that woke him up from sleep. The pain went away on its own, nothing made it worse.

Past medical history: hyperlipidemia

Family history: no history of heart disease

Social Hx: recreational marijuana use; remote history of cocaine and tobacco abuse (quit smoking 30 years ago). Does not exercise.

PHYSICAL EXAM

Vitals: T: 36.1 °C, HR 56, BP 138/88, RR 16, SpO₂: 97% on room air

General: muscular build, not in acute distress

Cardiac: normal S1/S2 cardiac sounds with no murmurs, no chest pain on palpation

Pulmonary: clear breath sounds bilaterally

Abdominal: no abdominal tenderness and normoactive bowel sounds

Musculoskeletal: full strength and sensation in extremities

Neuro: no focal deficits

Skin: no evidence of rash or petechiae

EKG: Abnormal T-waves in anterolateral leads. Minimal ST elevation in inferior leads.

Echocardiogram: anterior wall motion abnormalities consistent with the distribution of the left anterior descending artery

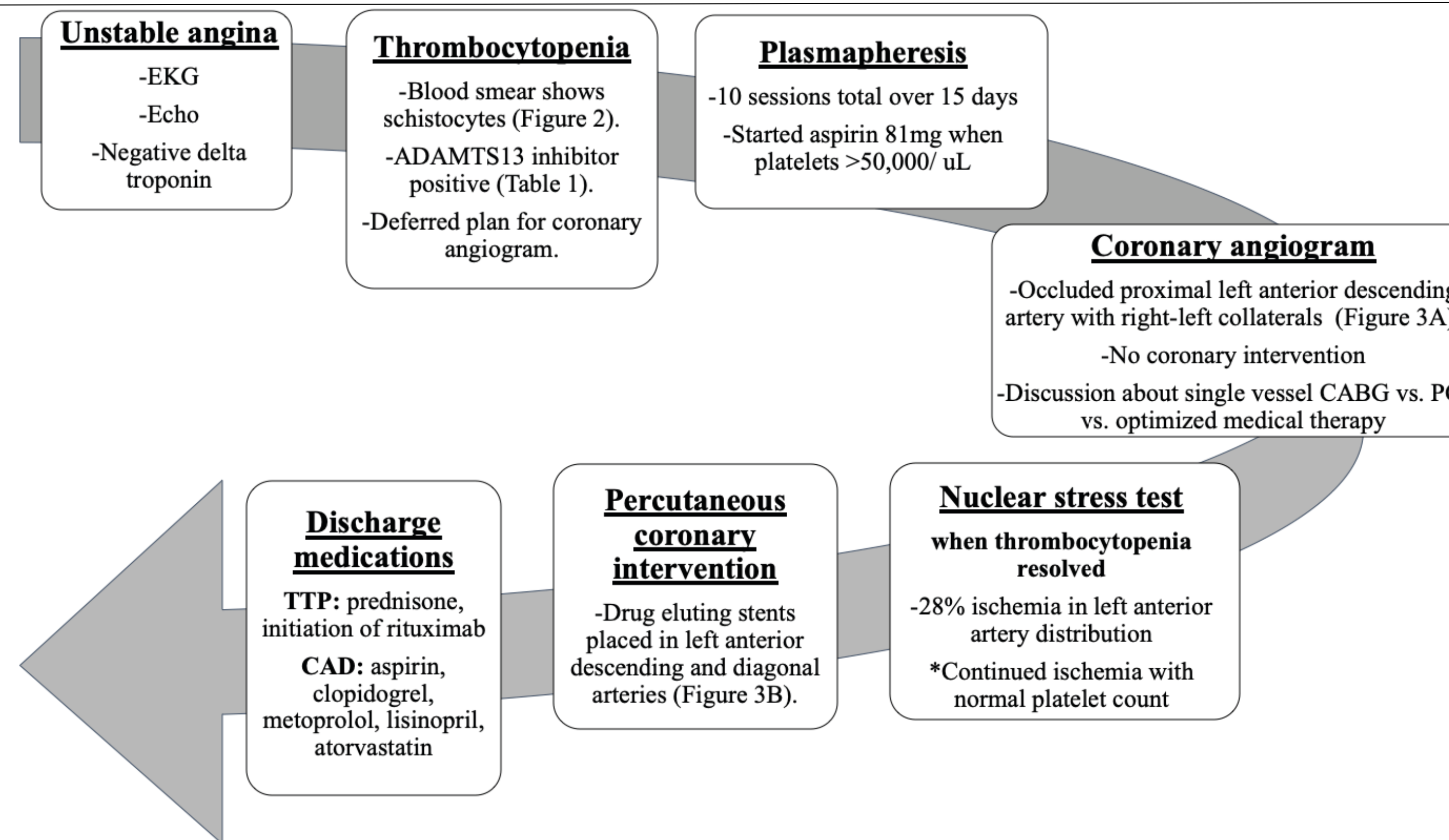


Figure 1: Clinical decision making course "

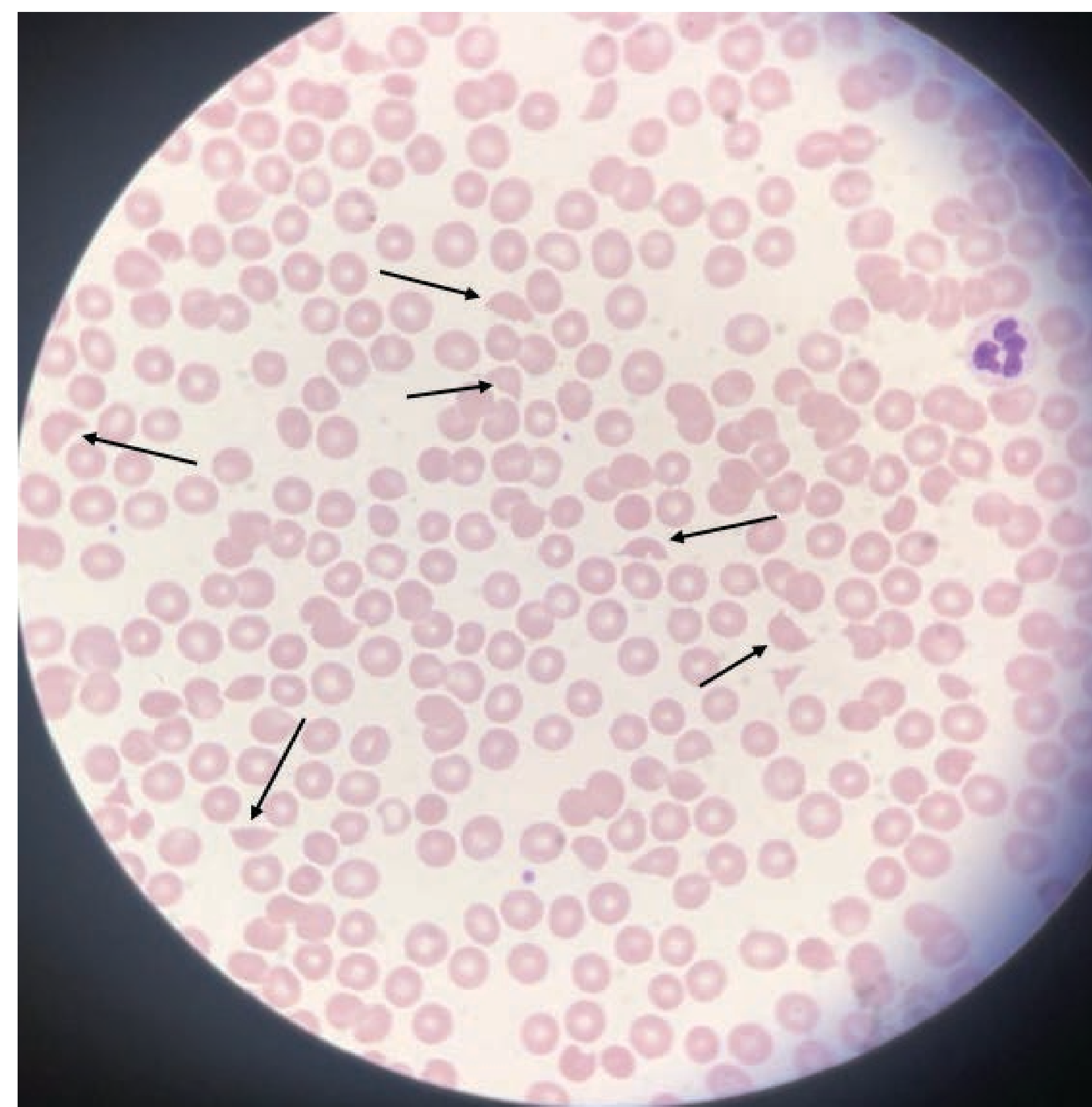


Figure 2: Peripheral smear with at least 6 schistocytes (arrows) per high power field.

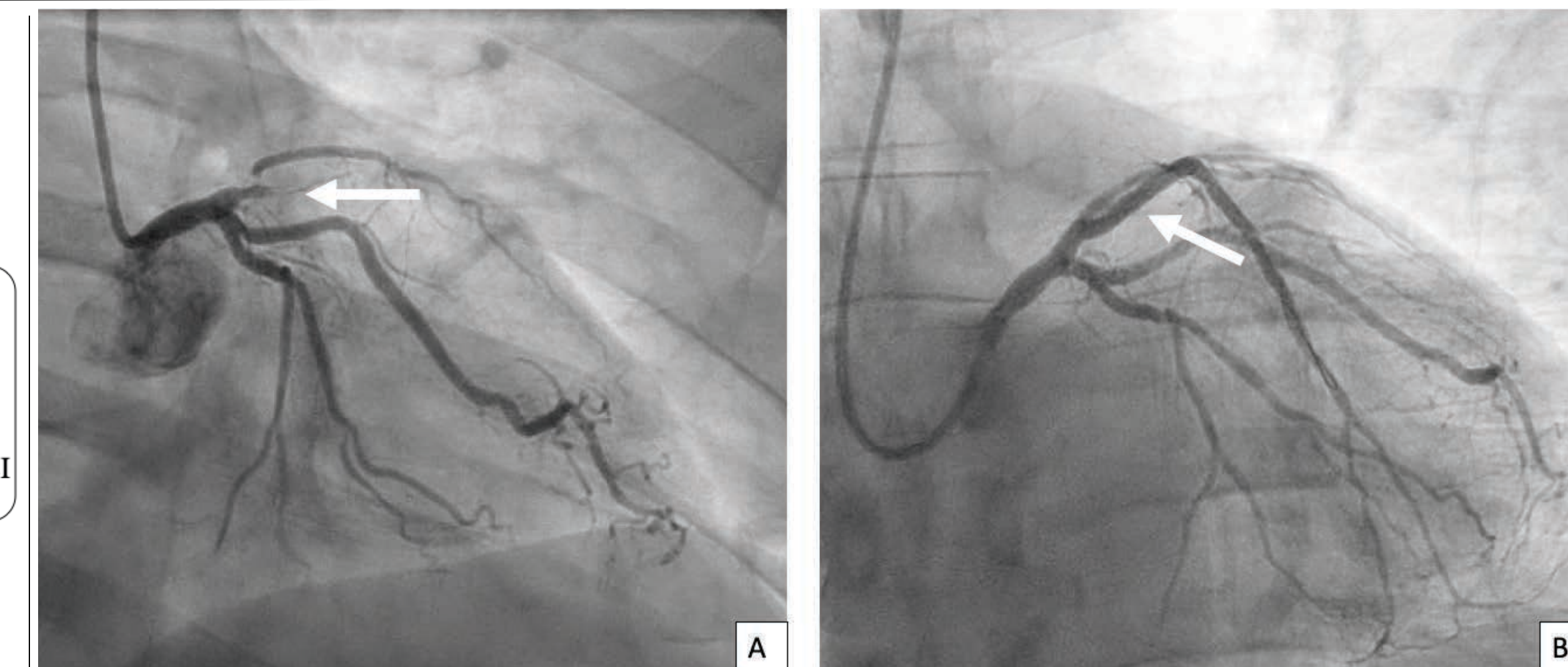


Figure 3. Coronary angiography, percutaneous coronary intervention. A. Occluded proximal LAD (white arrow) with right to left collaterals. B. Coronary arteries after percutaneous coronary intervention. Left anterior descending artery revascularized (white arrow).

Table 1. Pertinent Lab Results, abnormal results are in blue

Lab test \$	Value	Normal
0 hour troponin	14	0-21 ng/L &
3 hour troponin	18	0-21 ng/L &
Delta troponin	4 &	0-11
White blood cell count	5,600 / uL &	4,200-9,100 / uL
Hemoglobin "	12.3 g/dL	13.7-17.5 g/dL
Hematocrit	37%	40-51%
Platelets	20,000 / uL	150,000-330,000 / uL
Total bilirubin	3.2 mg/dL	0-1.2 mg/dL
Indirect bilirubin	2.8 mg/dL	0.1-1.0 mg/dL
Lactate dehydrogenase	643 U/L	118-225 U/L
Haptoglobin	<20 mg/dL	30-200 mg/dL
Prothrombin time (PT)	12.2 seconds	10-12.9 seconds &
International normalized ratio (INR) "	1.1	0.9-1.1
Activated partial thromboplastin time (aPTT)	31.6 seconds	25.8-37.9 seconds &
Fibrinogen	377 mg/dL	172-409 mg/dL &
D-dimer	2.08 ug/mL FEU	0-0.5 ug/mL FEU &
ADAMTS13 activity	6%	>70% &
ADAMTS13 inhibitor	Titer 1.6	<0.4

Conclusions

- Our patient had TTP and concomitant obstructive coronary artery disease.
- Maintain a broad differential for unstable angina, including TTP as a cause for chest pain, due to microthrombi in the coronary arteries.
- Before obtaining a coronary angiogram, platelets should be normalized, and there still has to be signs of ischemia.
- TTP affects the timing of angiography and initiation of antiplatelet medications.
- Starting antiplatelet medications is important for prevention of microthrombi formation.
- Rituximab is used to reduce the risk of exacerbation and relapse of TTP.

References

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