Case presentation
Acute kidney injury with red urine

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Clinical presentation

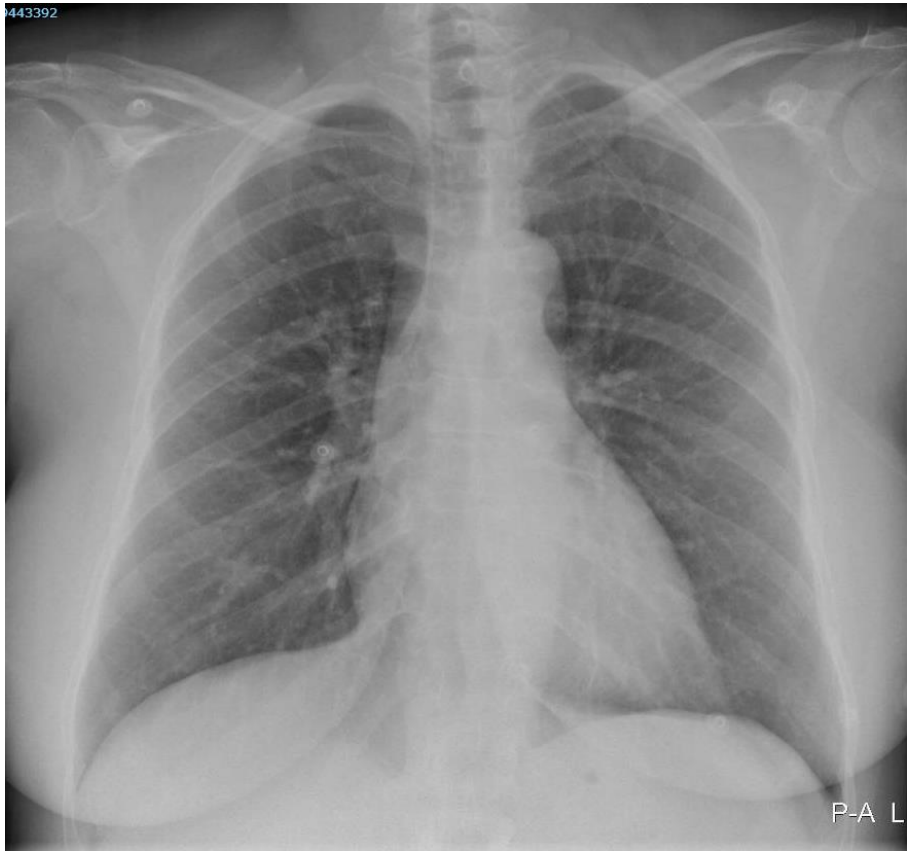
- 56 year old woman, Iranian descent
- History: RA, hypertension
- Medication: MTX 15 mg/wk, folic acid, celecoxib 2dd200mg, omeprazole 1dd20 mg, atenolol 1dd50 mg, HCT 1dd12.5mg
- ED: 1 day: malaise, nausea, vomiting, dyspnea and red urine, no flank pain
- Not acutely ill, alert, responsive. RR 155/90 mmHg, HR 90/min, T38.5, sat 92% (room air)
- Dry mucous membranes, normal breath sounds

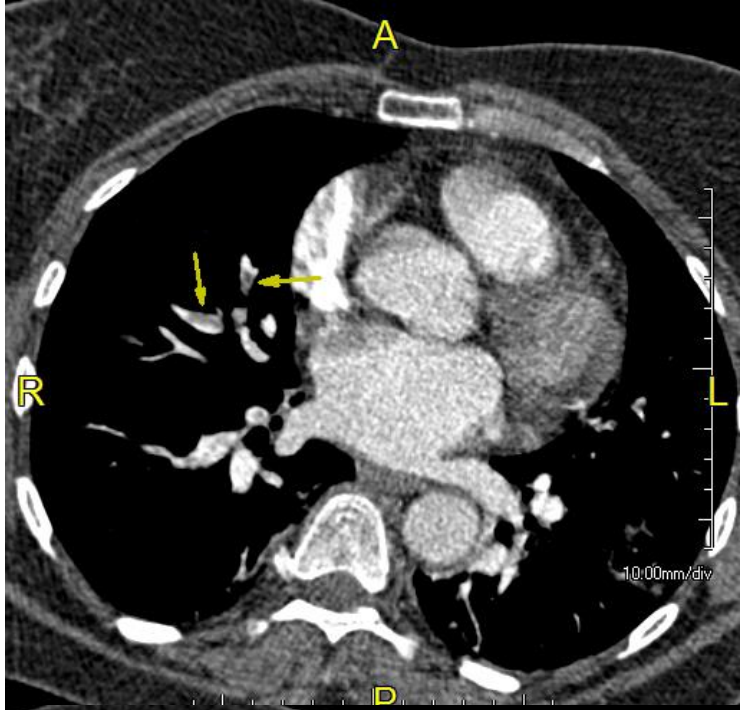
Laboratory results

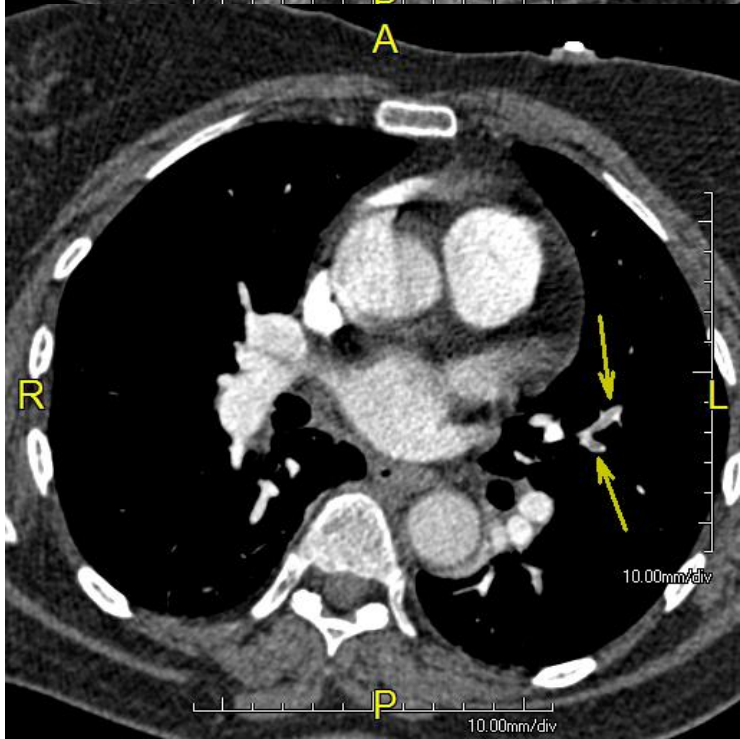
- Hb 5.0 mmol/L = 8.06 g/dL (3 months earlier 7.3 mmol/L), Ht 24%, L 16.0, T221, 4.6% reticulocytes
- Na 138, K 3.8, Creat 166 umol/L = 1.9 mg/dL (3 months earlier 66 umol/L), urea 21.6, CRP 31
- ALAT 67, ASAT 245, LDH 2390, AF 120, yGT 44, CK 155, tot bili 117 umol/L = 6.84 mg/dL, direct bili undetectable
- Haptoglobin undetectable
- Coombs test: negative

Laboratory results

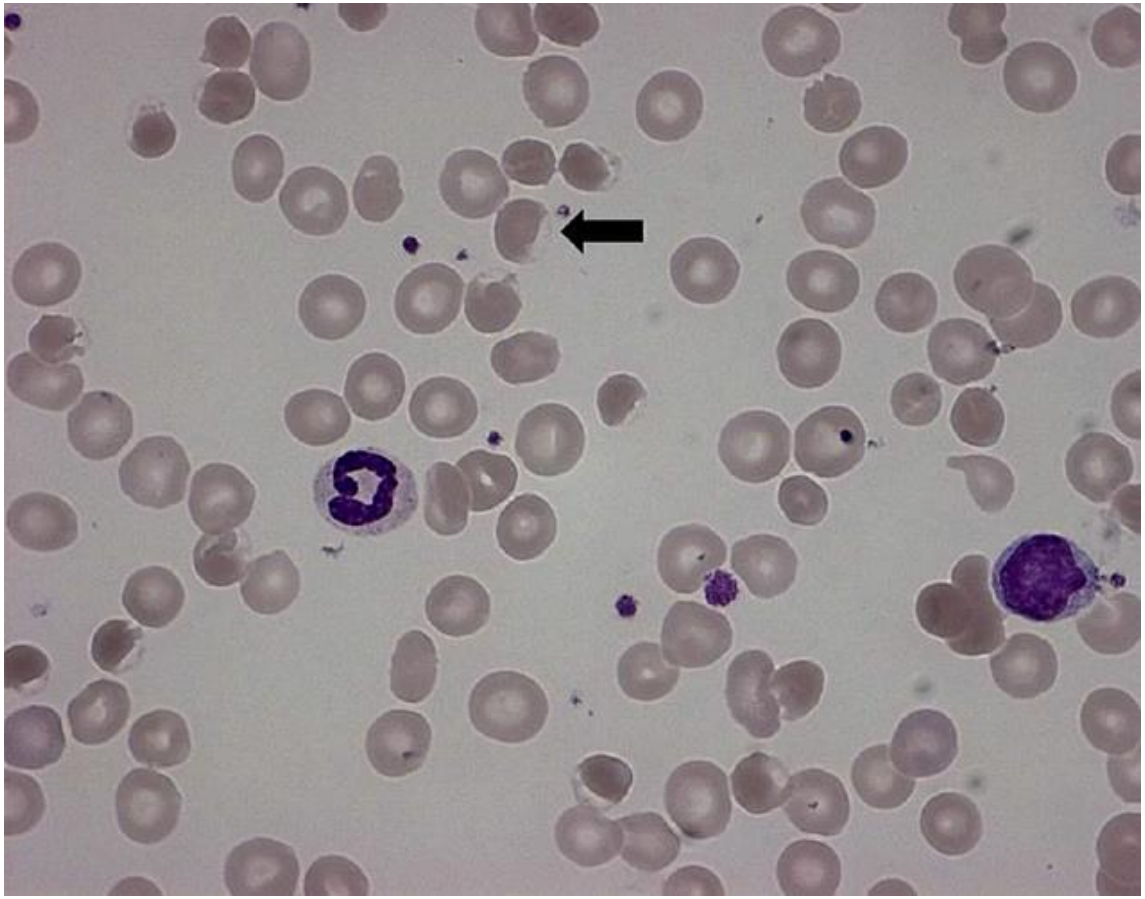
- Urineanalysis: haem (3+), protein (3+)
- Red supernatant
- Sediment: > 100 ery's, no dysmorphic cells nor acanthocytes, 5 leucocytes, 2-3 detached tubular cells, occasional granular casts
- Kidney ultrasound: unremarkable







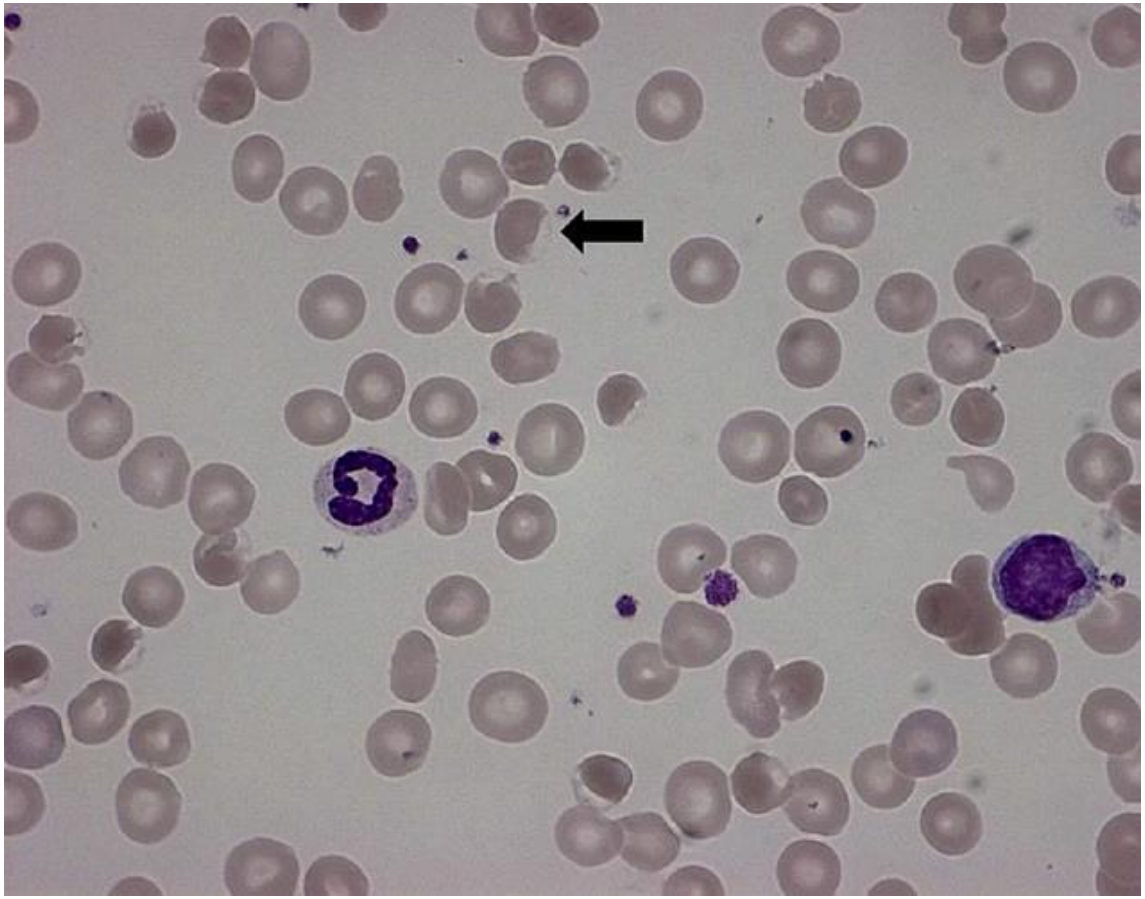
- ABGA: pH 7.47, pCO2 4.6 kPa (35mmHg), pO2 7.6 (57 mmHg)
- A-a gradient 6.7 (n<3)
- D-dimer: 39714 ng/mL
- Multiple pulmonary emboli



The blood smear helps!

Differential diagnosis of Coombs negative hemolysis and acute kidney injury:

- TTP or HUS?
- erythrocyte enzyme deficiency?
- Hemoglobinopathy?
- PNH?



- Blister cells and irregularly contracted cells (due to oxidative stress)
- Seen in G6PD deficiency
- Upon inquiry: 1 day before symptoms developed, she ate fava beans
- G6PD level proved undetectable (< 0,1 U/g Hb)

Questions

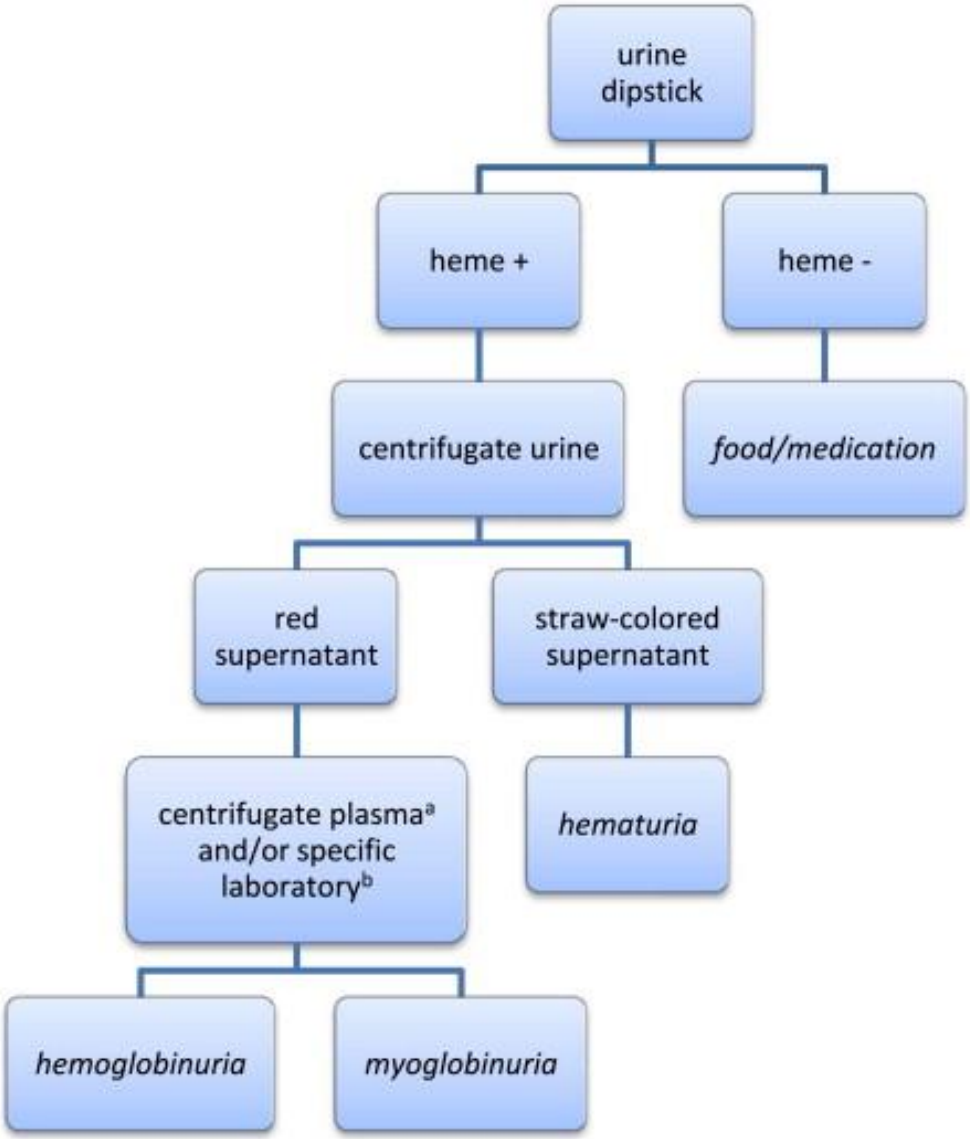
- Diagnosis: Favism complicated by AKI and pulmonary embolism.

1) What could be the mechanism of her pulmonary emboli and AKI?
2) What is the evaluation of red urine?
3) How should this patient be treated?

Mechanism PE and AKI

- Fava beans in G6PD deficiency → Massive intravascular hemolysis → free hemoglobin = potent NO scavenger → venous thrombosis and severe tubulotoxicity
- Dehydration (vomiting) combined with NSAIDs and diuretic → prerenal AKI + promotion PE
- Effect of iv contrast → AKI
- Inflammation (RA) → PE

Evaluation of red urine



How should the patient be treated?

- Pevention! Avoiding oxidative stressors
  - Fava beans
  - Infections
  - Oxidative drugs/ chemicals
    - Glibenclamide, co-trimoxazole, sulfazalasine
- Conservative treatment with fluid administration

Acetanilid
Diaminodiphenyl sulfone
Furazolidone (Furoxone)
Glibenclamide
Henna (Lawsone)
Isobutyl nitrite
Methylene Blue
Naphthalene
Niridazole (Ambilhar)
Nitrofurantoin (Furadantin)
Phenazopyridine (Pyridium)
Phenylhydrazine
Primaquine
Sulfacetamide
Sulfanilamide
Sulfapyridine
Thiazolesulfone
Trinitrotoluene (TNT)
Urate oxidase

Beutler et al, Blood 2008

Follow-up patient's renal function