ESIM 2016 Case Presentation

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Male, 36
Brought in by ambulance
Seen immediately upon arrival

Awake
ill-appearing
Profuse sweating
Icteric
Generalized petechial
hemorraghes/ecchymoses

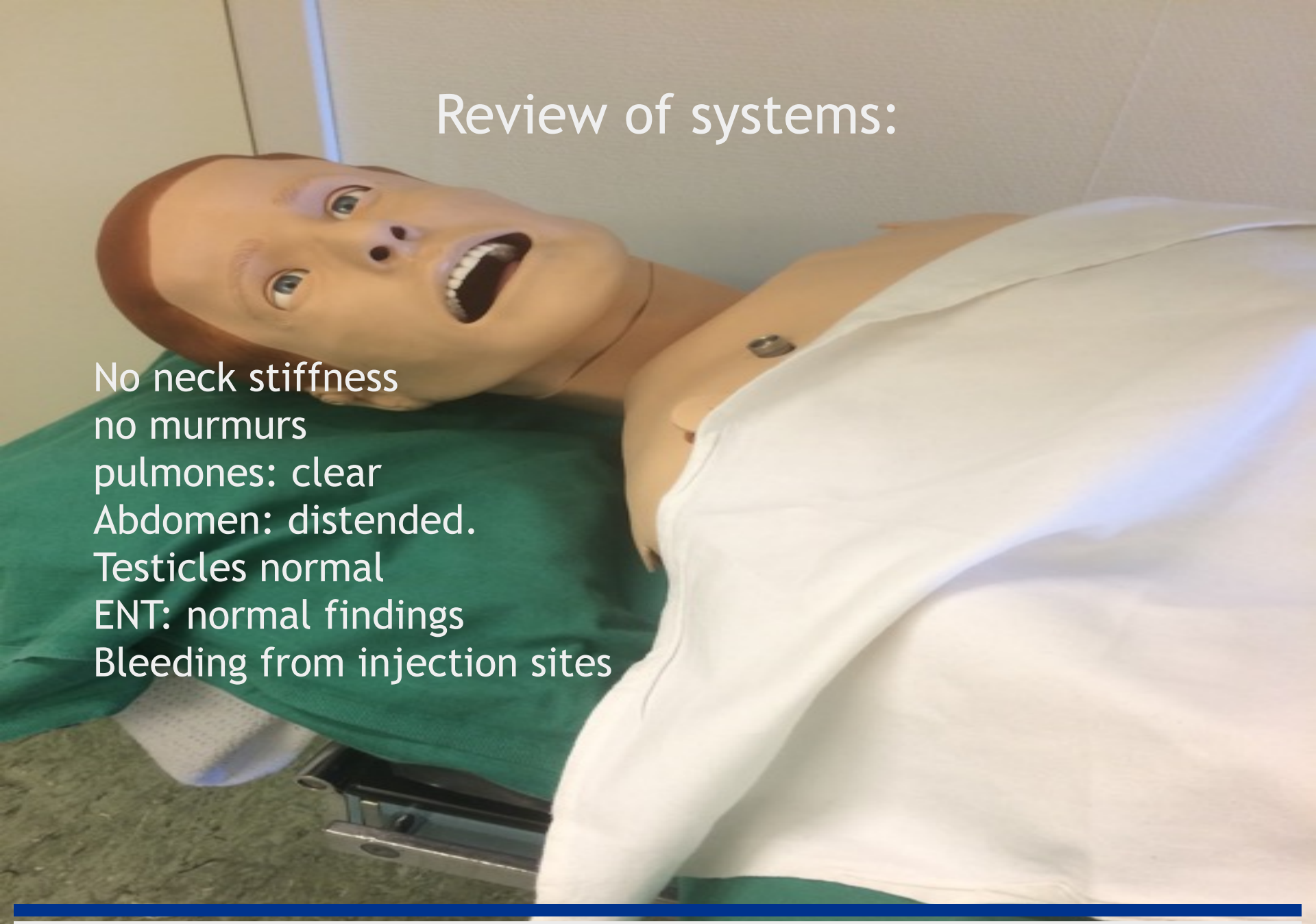
Vitals:
BP 118/50 mmHg
pulse 113 bpm, regular rhytm
Temperature: 40,1°C
Resp.rate: 24
Blood glucose: 13 mmol/L

Medical history

recently divorced, 2 small children
non-smoker, no substance abuse, no alcohol
no recent travels
no sick contacts
Hypothyroidism (Levothyroxine)
Mom with ‘immune-mediated’-disease

Abdominal/pelvic pain 4 months, neg. ultrasound
Gingival bleeding 4 weeks
Last 2 days: fatigue, chest pain, dyspnea, vomiting and noticing yellow discoloration of skin

Review of systems:



No neck stiffness
no murmurs
pulmones: clear
Abdomen: distended.
Testicles normal
ENT: normal findings
Bleeding from injection sites

Differential diagnosis?
Tests?

Arterial blood gas

pH 7.48
pCO2 2,8 kPa
pO2 23,6 kPa
HCO3 15 mmol/l
Base excess -7,8
Lactate 5,5

Hemoglobin 10,3 g/dl

EKG: diffuse ST-elevations in all precordial leads and inferior leads

Chest XR: clear.

Patient deteriorates in the ER, rushed to the intensive care unit

difficulties with iv-infusions, bleeding from skin, nose and mouth

No lumbar puncture performed

Antibiotics started
- ceftriaxone
- metronidazole

Patient is desoriented, GCS 13-14

Respiratory unstable, before intubation he becomes agitated and unconscious

Resp. arrest

Cardiac arrest. Advanced CPR started
(1 hr after arrival)

Lab is coming back:

Hb	10.1	(13.4-17 g/dl)
WBC	9.9	(3.5-10, x10^09/L)
Platelets	9	(145-390, x10^09/L)
CRP	19	(<5 mg/L)
Ddimer	10.2	(<0,5 mg/L)
Bili	82	(5-25 µmol/L)
Creatinine	265	(60-105 µmol/L)
TroponinT	1080	(<15 ng/L)

Advanced CPR is continued for 25 loops

One defibrillation despite PEA/asystole

Patient is declared dead about 2,5 hours after admission

Tentative diagnosis?

Autopsy performed next day

Multi-organ failure
Myocardial infarction and renal
Edematous shock-like organs
No spesific findings in lungs/liver/spleen
No signs of infection (blood cultures negative)

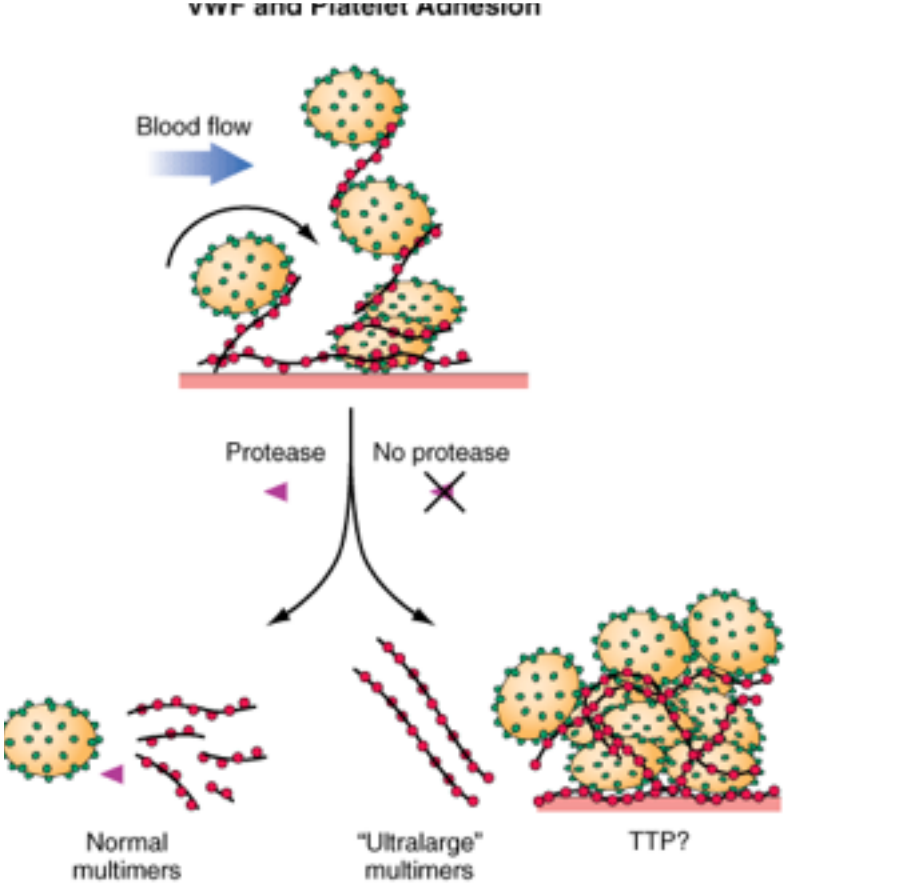
Brain
scattered thrombus formation. **Cerebellar tonsillar herniation (direct cause of death)**

Final diagnosis:

Thrombotic thrombocytopenic purpura (TTP)

1. Microangiopathic hemolytic anemia
2. Thrombocytopenia

- acute renal failure
- neurological symptoms (confusion, headache)
- fever



Source: Fauci AS, Kasper DL, Braunvald E, Hauser SL, Longo DL, Jameson JL, Loscalzo J
Harrison's Principles of Internal Medicine, 17th Edition: http://www.accessmedicine.com

often idiopathic

- medications, pregnancy, SLE, scleroderma

<u>ADAMTS-13-deficiency</u>:

→ protease that cleaves vonWillebrand factor

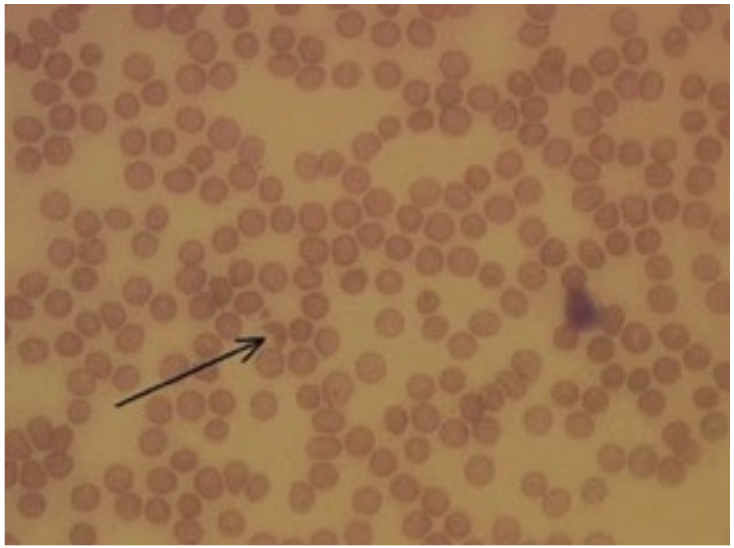
- congenital defect
- autoantibodies
- results in large WF-multimers; plateleg aggregation and thrombus formation

Microangiopathic hemolytic anemia

- Non-immune mediated hemolysis (neg. Coomb's-test)
- schistocytes in peripheral blood smear

Typical lab findings:

- LD↑
- Haptoglobin ↓↓
- Bilirubin ↑
- Normal INR/bleeding time



Schistocytes: mechanical stess upon erytrocytes due to to platelet aggregation

- Life threatening disorder
- Mortality ~ 90 % if untreated
- Plasmapheresis for antibody removal first line treatment
  - Glucocorticoids?
  - Rituximab?
  - Immunosuppressants?
  - Splenectomy