Cutaneous necrosis and destructive polyarthritis – a puzzle solved in 17 days

Radim Špaček Department of general internal medicine Hospital in Frýdek - Místek Czech Republic

64 year old man comes to the ER...

- History insignificant
- Complaints diarrhea for a few days, hematuria afterwards, then joint pains (mainly wrists, knees and ankles) and "rash".
- Physical exam
- K⁺ 2,84 mmol/l [3,5 5,3]
- Creatinine 140 µmol/l [40 100]
- <u>CRP 360 mg/l</u> [<5]
- WBC 15,7 x 10⁹/l [4 9 x 10⁹]



First differential??

- Hemorrhagic cystitis with reactive polyarthritis and erythema nodosum?
- Cryptogenic sepsis?
- Vasculitis?

Starting treatment – empiric ATB, analgetics,
K+ supplementation, iv. fluids

Moving the patient to the ICU ...

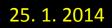
• Excrutiating joint pain, dyspnea, CRP still high

 Imipenem/Cilastatin, steroids, searching for infection negative repeatedly

Newly forming ulcers!







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New differential?

- Infection less likely, maybe TB?
- Vasculitis?
- Paraneoplastic syndrome?
- Plague? 🙂

• Turning point ahead...

PPP syndrome

Pancreatitis, polyarthritis, panniculitis

- Extremely rare (1970 2008 about 30 case reports worldwide)
- Etiology and pathogenesis acute or chronic pancreatitis, pseudocysts, tumors (adenocarcinoma from acinar cells)





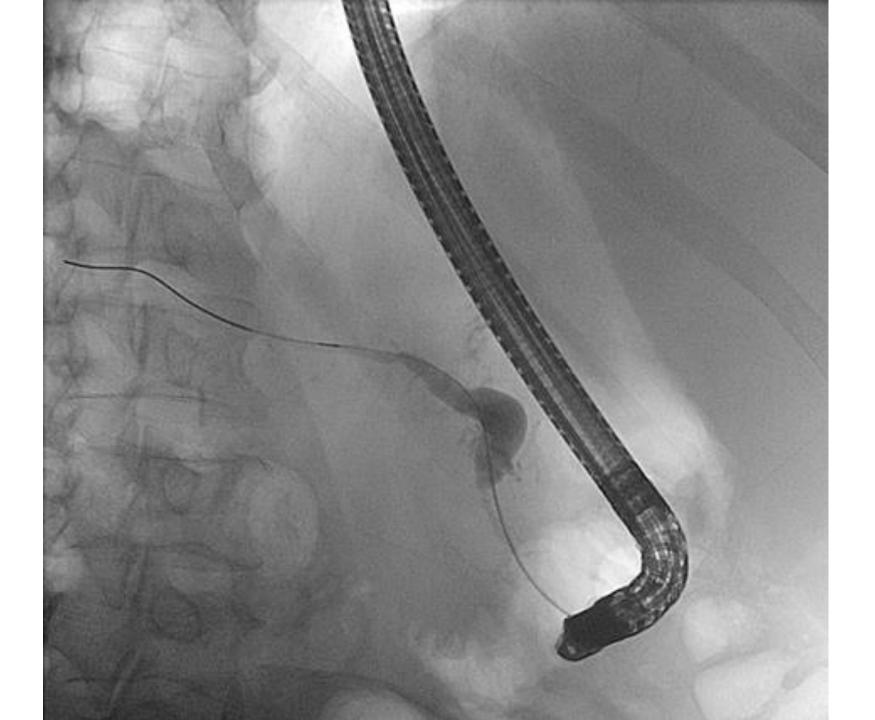
Confirming the diagnosis ...

- Blood chemistry: AMS 21, LPS 68 μkat/l [<2]
- Synovial fluid: AMS 7, LPS 9 μkat/l
- Cutaneous biopsy from lower limb: fat necrosis

Treatment and aftermath

- Combined enteral and parenteral nutrition
- High-dose Octreotide
- Endosonography
- ERCP

Beskydske gastrocentrum ID: OLYMPUS NAME: AGE: DOB: SEX: 18/02/2014 13:32:43 6MHz 9cm G:17/19 I:L1 c:3/8 FC:1 L.DEN:x1.0 ±6.0 TX: 88% CFG: MEDIA 19 T/B:ROI LOC DIR: ----NOR THE PERSON SCL: CNCT:L R 10mm





Aftermath ...

- Patient moved to a surgical ward
- Despite an intensive wound care, he subsequently needed amputation of both his legs
- Right arm was endangered, but saved
- Moved to rehabilitation facility after 11 months in hospital

Take home messages

- More common diseases are more common, but rare ones exist as well
- Erythema nodosum in older patients!
- Measure AMS and LPS in unusually presenting arthritis













