ESIM winterschool 2012

Clinical Case Presentation

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initial presentation

35-year old female patient

admitted to the hospital in suspicion of pyelonephritis

- back pain for a week
- now flank pain
- shivering, nausea, and vomiting
- anuria since the day before

PMHx: none

Medication: none

Social Hx:

- Cuban, living in Germany for 2 years now
- married, no children

Physical examination

Main findings:

neurology:

paraparesis of legs with reduced sensory function

abdomen:

bowel sounds decreased

pressure pain in the lower abdomen

full bladder

additional findings:

general aspect:

reduced

normal weight (56kg, 163cm, BMI 21kg/m^2)

no fever (37,6°C)

cardiorespiratory:

stable (P 75/min, RR 130/75)

Laboratory results

- **HGB** (12.3-15.3): 12.6 g/dl
- **PLT** (130-350): 254.6 g/l

- **CK** (0-170): **12,000 U/l;** CKMB (7-25): **329 U/l**

- **Na** (135-147): 142 mmol/l
- **CRP** (< 5): 6.3 mg/l
- **Crea** (0.6-1.0): 0.69 mg/dl

- **elektrophoresis**:
  IgG-gammopathy (700-1600mg/dl): 1790.0

- **urinalysis:** without pathological findings

Key findings up to this point????????

main diagnostic hypothesis

Suggestions???????????

dd: paraparesis

dd: urinary retention

dd: ck increase

Additional diagnostic work-up (I): laboratory studies

- **C3** (90-180): 107 mg/dl
- **C4** (10-40): 13.4 mg/dl

- **cANCA**: neg.
- **pANCA**: neg.

- **ENAS**: positive
- **ENAP**: SSA-AK

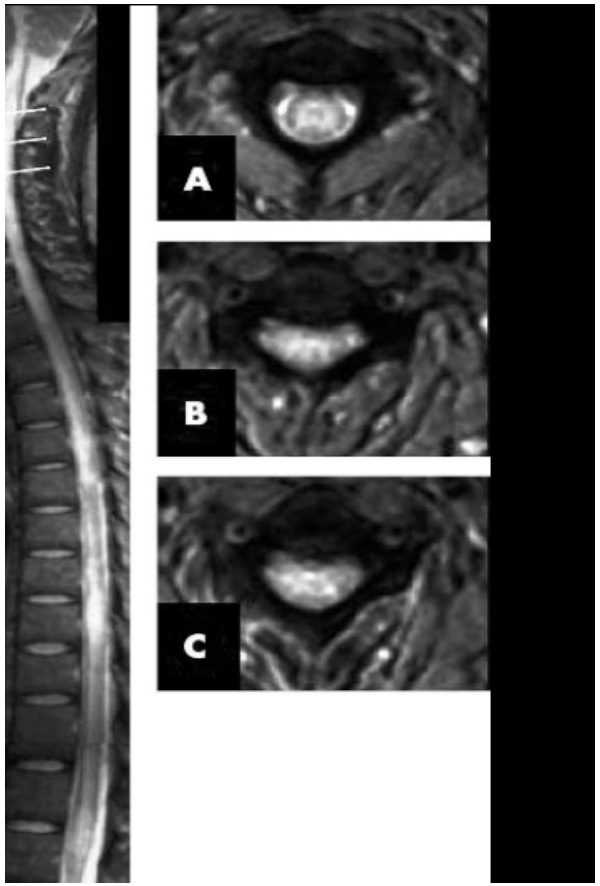
serologic testing:

- Syphillis (pox): neg.
- BorrlgG (borreliosis): neg.
- HBsAK: neg.
- HCV AK: neg.
- HIV: neg.

Additional diagnostic work-up (II):

MRI: T2 showed multiple and confluent high signal intensities within the spinal cord; diffuse swelling

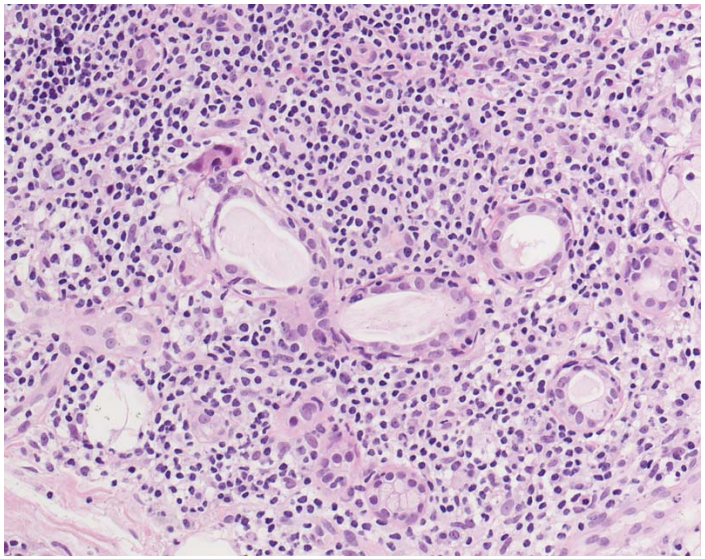
-> myelitis C2 – TH1



T Yamamoto, S Ito, T Hattari (2006)
Acute longitudial myelitis as the initial manifestation of SS; J Neural Neurosurg Psychiatry; 77:780

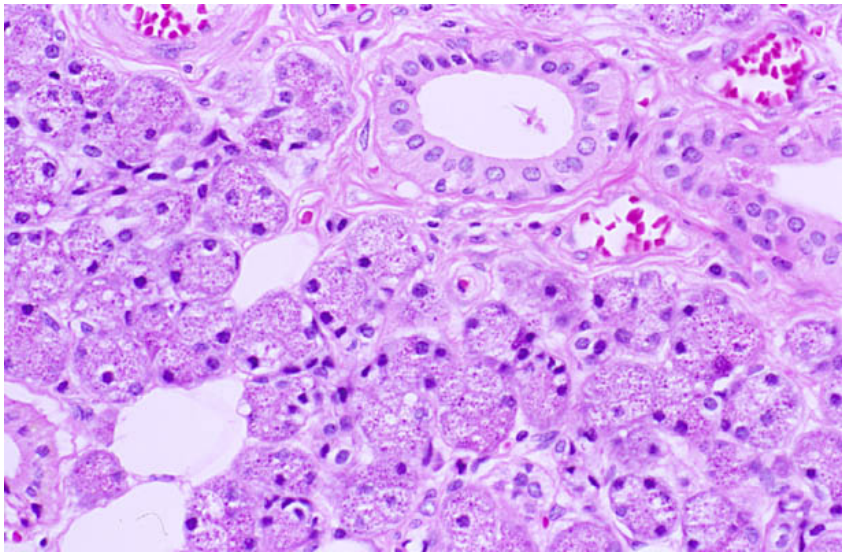
Additional diagnostic work-up (III):

- Salivary gland biopsy: focal lymphocytic sialadenitis,
  (50 or more lymphocytes per 4 mm²)



http://alf3.urz.unibas.ch/pathopic/getpic-img.cfm?id=8280

normal parotid histology



http://pathology.mc.duke.edu/website/images/grad/Histo_course/parotid.jpg

diagnosis: **Sjogren-Syndrome**

American-European Classification Criteria (09/2011)

- Ocular symptoms of inadequate tear production
- Ocular signs of corneal damage due to inadequate tearing
- Oral symptoms of decreased saliva production
- Salivary gland histopathology demonstrating foci of lymphocytes
- Test indicating impaired salivary gland function
- Presence of autoantibodies (anti-Ro/SSA and/or anti-La/SSB)

Course of disease & treatment approaches

-> progression of hemiplegia during first 3 days after admission

- high-dose corticosteroid therapy for 5 days (1g/d)

-> aggravation: onset of tetraplegia & respiratory insufficiency causing need for intubation/ BIPAP-ventilation

- switch to an adjunction:

non-steroidal immunosuppressant: cyclophosphamide 1 g every 3 weeks
plus
corticosteroids (1000 mg/d)

-> decrease of ck-levels to normal, but still neurological symptoms

- therapeutic stepping up:

high dose cyclophosphamide (30g) for 5 days

-> the patient was able to move her right arm after a week of treatment

Therapeutic alternatives in literature:

fulminant development ----→ application of rituximab indicated?????

Follow-up

- *4-6 weeks after onset of treatment:*

  the patient was slowly able to move her right side and left arm

- consecutively CPAP weaning possible,
  after 3 months of hospital stay: effective extubation, transfer to rehabilitation

- *resulting long-term damage*:
  - hemiplegia (left)
  - sight disorder

- CAVE: patients with Sjögren syndrome have a higher rate of B-cell lymphoma (6-8 fold risk)

Key learning points

case complexity: medical benefits from an interdisciplinary team play

Idealistic view or reality in your daily working life??????

atypical manifestation of illnesses: a challenge for the detective in each of us

References

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