Juvenile-oncet Systemic Lupus Erythematosus with neuropsychiatric manifestations: a case report

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Background

Juvenile-oncet SLE (in comparison with SLE in adults):
- More severe
- More often CNS involvement [1][2]
  - CVD caused by thrombosis
  - Syndromes caused by systemic inflammation
    - demyelinating syndrome, headache, aseptic meningitis, chorea, seizures
    - psySLE: anxiety disorder, acute confusional state, cognitive disorder, mood disorder, psychosis.

Diagnostic problems
- Lack of "golden" diagnostic standards
- Headache, mood disorders, mild cognitive disorders are common in JSLE [3]
- Differential diagnosis (corticosteroid-induced psychiatric disease) [4]

Patient presentation

- 17 y.o. girl after a suicide attempt (multiple cut wounds of forearms)
- Before the episode:
  - Depressive illness ≈ 3 years
  - Photosensitive skin rash ≈ 2 years
  - Low extremities pain ≈ 1 years
  - mild hypochromic anaemia, tendency to leucopenia($4{,}0 \times 10^9$) ≈ 6 months
- post-hemorrhagic anemia, leukopenia (2.3×10^9), neutropenia (0.7×10^9),
- increased RF (211 IU/ml). ANA positive (1:2560)
- anti-dsDNA, aPL: negative.
- Synovitis of knee joints confirmed by US
- Cranial MRI: subcortical FLAIR hyperintensity in the right frontal lobe (unspesific) (Figure 1), with normal EEG

The diagnosis of
jSLE with neuropsychiatric involvement
was suggested

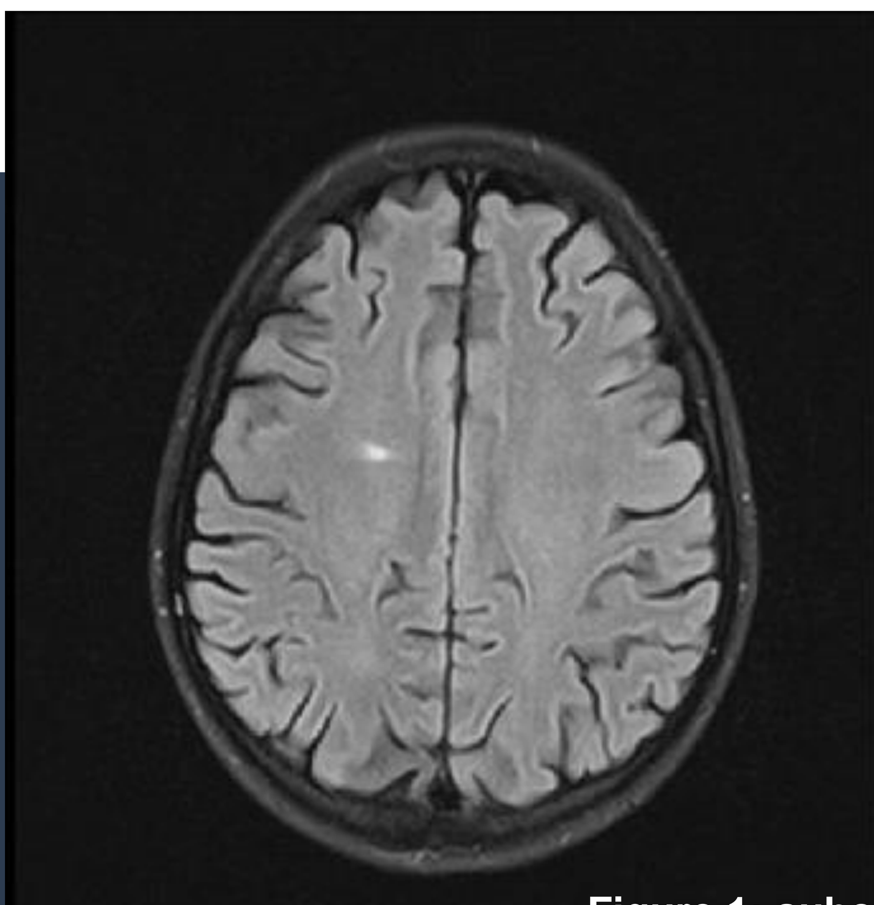
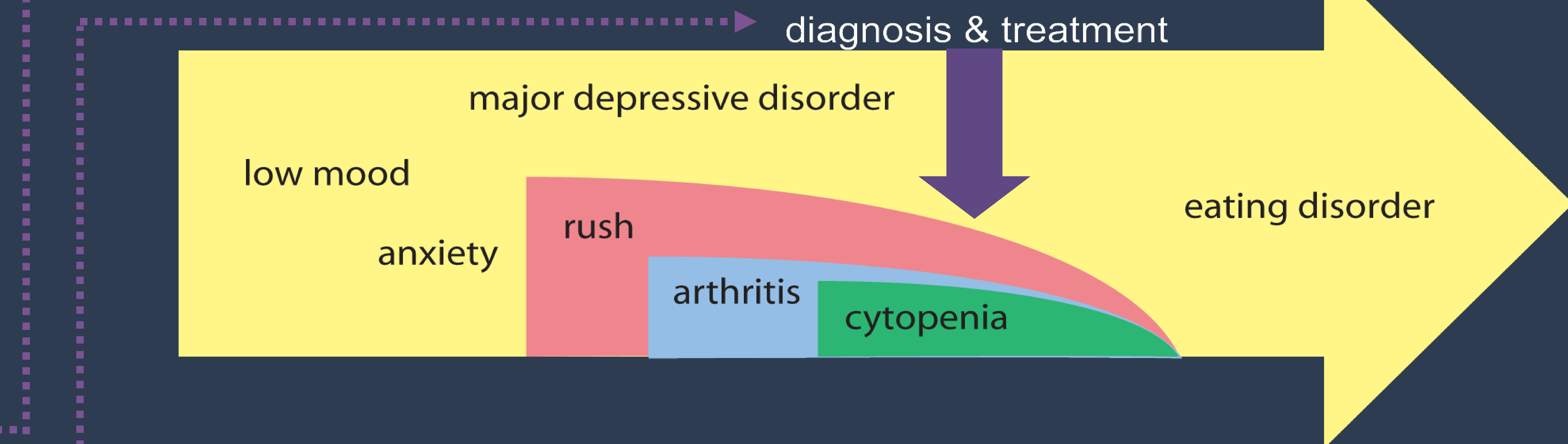


Figure 1: subcortical FLAIR hyperintensity in the right frontal lobe on cranial magnetic resonance imaging (MRI). Such findings are common for NL, but unspecific [4]

Timeline



Interventions

- **immunosuppressive therapy:**
  - intravenous cyclophosphamide 700 mg/m2/month (№3),
  - intravenous methylprednisolone pulse (№3) therapy followed by
  - oral prednisone (1,4 mg/kg with further tapering)
  - hydroxychloroquine.
- **Antidepressant (Fluoxetine)**
- **Anxiolytic (Hydroxyzine)**
- **Psychotherapy**

Results

+ normalization of blood tests
+ improvement of arthritis
+ improvement of skin rash
+ significant emotional improvement

- continued eating disorder (poor control, life-threatening)

Discussion

Was it neurolupus initially?

+ significant improvement after immunosupressive therapy
- NL is associated with generally severe disease course

Major depressive disorder:
- not included in classification criteria (ACR-97, SLICC, EULAR/ACR-2019)
- not included in activity score systems (SLEDAI, ECLAM), but:
  - included in SLAM (severe depression)
  - included in BILAG (organic depressive illness)

Further improvement of our diagnostic instruments is needed

Conclusion

Psychiatric disease can mask not only psySLE but any developing somatic disorder.

- organic psychiatric disease tends to become persistent without appropriate treatment
- the interdisciplinary approach (cooperation of psychiatrists, rheumatologists and neurologists) is necessary, especially in atypical cases

Acknowledgements, references, contact information

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List of abbreviations:
CNS – central nervous system
CVD – cerebrovascular disease
NL – neurolupus
psySLE – systemic lupus erythematosus with psychiatric manifestations
SLE - systemic lupus erythematosus
jSLE – juvenile-oncet SLE