Differential diagnosis of children with symptoms suggestive of transverse myelitis

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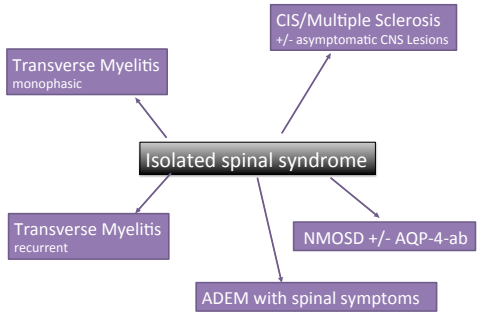
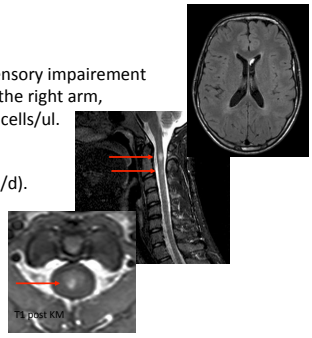
Differential diagnosis

1. immune mediated
   isolated spinale syndrome (Transverse myelitis, NMO...)
   ADEM
   systemic autoimmune diseases (e.g. SLE)
2. infektious
   viral: EBV, Coxsackie A/B, HIV, HSV, VZV,...
   bacteriel: Borrelia burgdorferie
3. paraneoplastic (Glioblastome, ependymome...)



Case 1

- 14 yo boy with sensory impairement and weakness in the right arm, CSF: OCB pos, 23 cells/ul.
- Remission after IVMP (20mg/kg 3/d).
- Clinically stable after 18 month.



Diagnosis MS – *MRT criteria*

TABLE 1: 2010 McDonald MRI Criteria for Demonstration of DIS

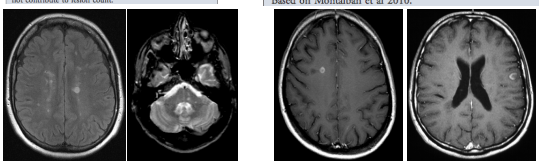
DIS Can Be Demonstrated by ≥1 T2 Lesion[a] in at Least 2 of 4 Areas of the CNS:
Periventricular
Juxtacortical
Infratentorial
Spinal cord[b]

Based on Swanton et al 2006, 2007.[22,27]
[a]Gadolinium enhancement of lesions is not required for DIS.
[b]If a subject has a brainstem or spinal cord syndrome, the symptomatic lesions are excluded from the Criteria and do not contribute to lesion count.

TABLE 2: 2010 McDonald MRI Criteria for Demonstration of DIT

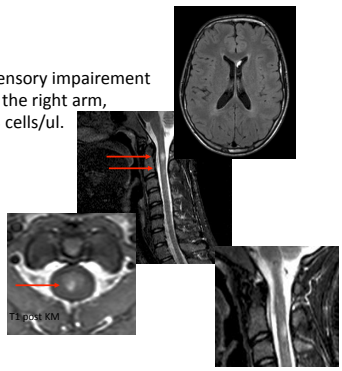
DIT Can Be Demonstrated by:
1. A new T2 and/or gadolinium-enhancing lesion(s) on follow-up MRI, with reference to a baseline scan, irrespective of the timing of the baseline MRI
2. Simultaneous presence of asymptomatic gadolinium-enhancing and nonenhancing lesions at any time

Based on Montalban et al 2010.[24]



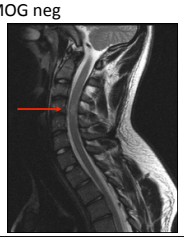
Case 1: CIS

- 14 yo boy with sensory impairement and weakness in the right arm, CSF: OCB pos, 23 cells/ul.
- Clinically stable after 18 month.



Case 2: monophasic transverse myelitis

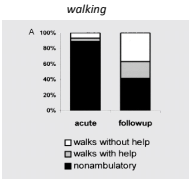
- ✓ 8 yo girl with tingling in both legs worsening over 24 hours, no pain;
- ✓ Neuro-Exam: incomplete sensory level C4 (decreased sensation for light touch and cold/warm, no weakness
- ✓ CSF: 83 cells/µl, OCB neg, AQP4/MOG neg
- ✓ Tx: IVMP for 5 days
- ✓ 2 y-FU: normal neurological exam, cMRI normal.



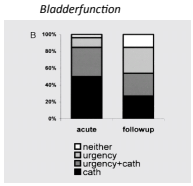
Transverse Myelitis

- Bimodale age distribution: <3y and 5-17y
- M:f 1.04
- 42/47 cases (89%): rec TM, ADEM, NMO, MS

walking



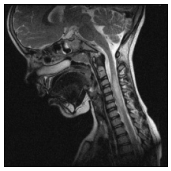
Bladderfunction

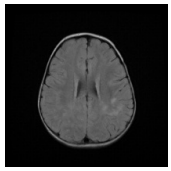


Pidcock et al. Neurology 2007; 68: 1474-1480
n=47, retrospective, follow-up: 8y (CI: 4.5-11.9)

Case 3: AQP4pos-NMOSD

- ✓ 12 yo boy with an episode of hiccups and weakness in his legs
- ✓ CSF 120 cells/µl, no additional OCBs

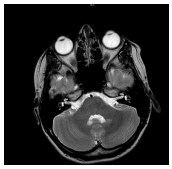
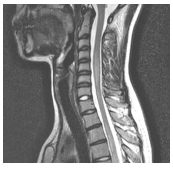




- ✓ Longitudinale Extensive Transverse Myelitis (LETM)
- ✓ AQP4-IgG Abs present

Case 4: AQP4negNMOSD with MOGabs

- 12/2009: Weakness in both legs, bladder dysfunction
- 06/2010: Visual loss right eye
- MRT-c: cerebellar and myelon lesions spanning > 3 segments
- AQP4-abs absent

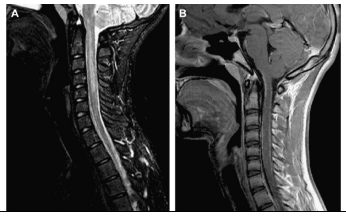
✓ Diagnosis: Neuromyelitis optica spectrum disorder

✓ High MOG-abs titer present!!!!

(Rostasy et al., MSJ 2013).

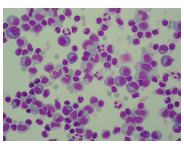
Case 5: Neuroborreliosis

- 16 yo boy developed a bilateral weakness in arms and legs, he has no pain, no bladder dysfunction.
- Spinal MRI: longitudinally extentive transverse myelitis (LETM), meningeale enhancement!
- CSF: Protein 109mg/dl, 442 cells/µl.



Case 5: Neuroborreliose

- Spinal MRI: longitudinally extentive transverse myelitis (LETM), meningeale enhancement!
- CSF: Protein 109mg/dl, 442 cells/µl.
- IgM und IgG antibodies against Borrelien present
- Borrelia burgdorferi-IgG- Antibody index (AI) elevated!

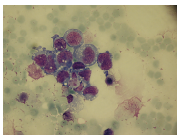


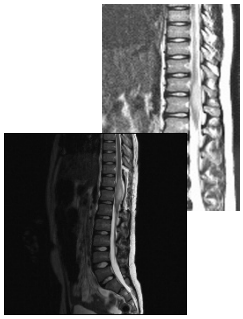
Case 6: Glioblastoma

- 12 yo girl with weakness left foot and severe back pain, CSF: 88 cells/µl.

✓ Dx: transverse myelitis

✓ 3 weeks later backpain and weakness in left foot.





Main points for the clinical practice (1) Transverse Myelitis

➢ Typical for CIS/ MS
- develops within hours
- Incomplette myelitis
- often only sensible deficits
- Lhermitte sign positive
- improvement without treatment

➢ Untypical for MS
- slow development of symptoms
- complete myelitis
- areflexia
- no spontaenous improvement
- pain

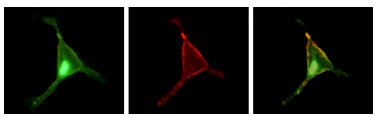
Main points for the clinical practice (2)

✓ MS/CIS: lesions are predominately in the cervical myelon and are associated with mild symptoms.

✓ If no persting improvement with steroids think of alternative diagnosis

✓ Pain in transverse myelits is rare!

✓ MOG antibodies are often present in children with LETM



Role of cerebrospinal fluid analysis in children with CNS infections/ encephalitis/vasculitis in the context of immunsuppression
to be added