Acute visual loss

INSELSPITAL
UNIVERSITÄTSSPITAL BERN
HOPITAL UNIVERSITAIRE DE BERNE
BERN UNIVERSITY HOSPITAL

Maja Steinlin

NEUROZENTRUM
Neurocentre | Neurocentro
Inselspital Universitätsspital Bern
Universitäre Psychiatrische Dienste Bern
Neurochirurgie
Neurologie
Neuropädiatrie
Neuroradiologie
Psychiatrie

KINDERKLINIKEN Bern

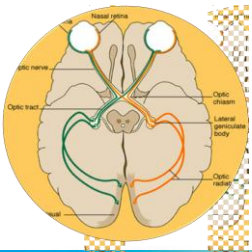
Sandra, 14 year old

- Previously healthy
- 10 day hx of worsening vision of the right eye

Sandra, 14 year old

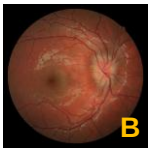
- Previously healthy
- 10 day hx of worsening vision of the right eye

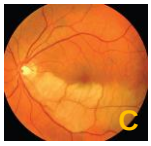
Where is the problem?



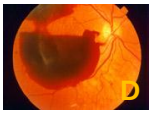
Differential possibilities?

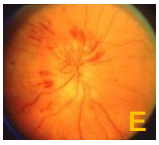
Most important clinical examinations?

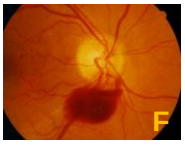


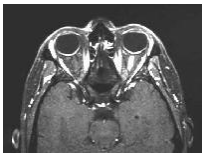
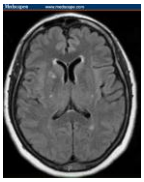
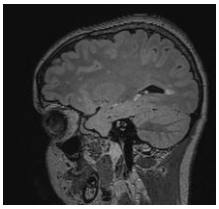


What are the diagnosis?
- all but one have visual problems







Differential diagnosis of optic neuritis

- Parainfectious optic neuritis
- Neuromyelitis optica Devic disease
  MAG or Aquaproin antibodies postive
  recurrent optic neuritis – optimal ttm wth Rituximab
- Multiple sclerosis
- Systemic disorders (as Sarcoidosis, Vasculitis, Lupus, Syphilis)
- (Leber's hereditary optic neuropathy)

Typical suggestive for MS versus Atypical for all the others

	Typical	Atypical
Age	Young adult	Age >50 years or <12 years
Ethnic origin	White	African, Asian, or Polynesian descent
Laterality	Unilateral symptoms	Bilateral simultaneous or rapidly sequential
Pain	Mild periocular pain; worse on eye movement	Severe periocular pain waking patient from sleep, painless visual loss, pain persisting longer than 2 weeks
Vision	Mild to moderated uniocular visual loss followed by spontaneous improvement	Severe visual loss (worse than 6/60 or 20/200), no recovery starting within 3 weeks of onset, progression of visual loss for more than 2 weeks
Appearance	Normal or swollen optic disc	Severe optic disc swelling, macular star (neuroretinitis), optic disc haemorrhages, anterior—posterior segment inflammation, marked retinal exudates
Other	Uhthoff's phenomenon, Pulfrich effect, previous self-limiting neurological episodes	Family history, neoplastic history

Table 2: **Features of typical and atypical optic neuritis**

Hints for special differentials

- Parainfectious optic neuritis
  - Children < 12 years,
  - bilateral optic neuritis
  - Infection within the last few weeks
- Neuromyelitis optica Devic disease
  MAG or Aquaproin antibodies postive
  recurrent optic neuritis – optimal ttm wth Rituximab
- Multiple sclerosis
- Systemic disorders (as Sarcoidosis, Vasculitis, Lupus, Syphilis)
- Leber's hereditary optic neuritis

Hints for special differentials

- Parainfectious optic neuritis
- Neuromyelitis optica Devic disease
  MAG or Aquaproin antibodies postive
  recurrent optic neuritis
  ttm with Azathioprin/Steroids second line Rituximab
- Multiple sclerosis
- Systemic disorders (as Sarcoidosis, Vasculitis, Lupus, Syphilis)
- Leber's hereditary optic neuropathy

Table 1
Comparison of typical features of childhood neuromyelitis optica and multiple sclerosis

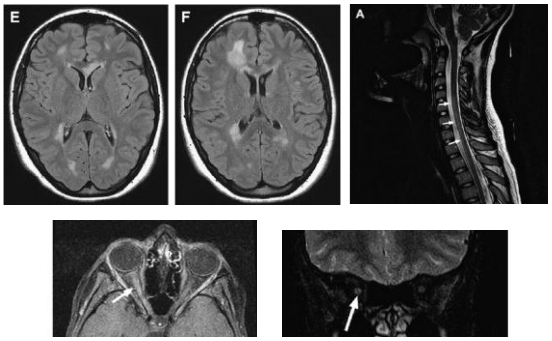
	Neuromyelitis Optica	Multiple Sclersosis
Female:male ratio	Up to 7:1	2:1
Relapsing course	53%–100%	96%
Secondary progression	Rare	Common
CSF characteristics	Marked pleocytosis (often >50 cells/μL) Lymphocytes or neutrophils	Mild pleocytosis (typically <30 cells/μL) Lymphocytes
CSF oligoclonal bands	90%	
MR imaging brain	Normal, nonspecific white changes, or characteristic diencephalic and brainstem lesions	
MR imaging spine	≥3 segments with central/holocordinvolvement	

Box 2
Diagnostic criteria for pediatric neuromyelitis optica

- Optic neuritis
- Transverse myelitis
- One of the following:
  - Longitudinally extensive spinal lesion (≥3 spinal segments)
  - NMO IgG seropositivity

Adapted from Krupp LB, Banwell B, Tenembaum S. Consensus definitions proposed for pediatric multiple sclerosis and related disorders. Neurology 2007;68:S7–12.

 Makhani et al, Neuroimag N Am 2013

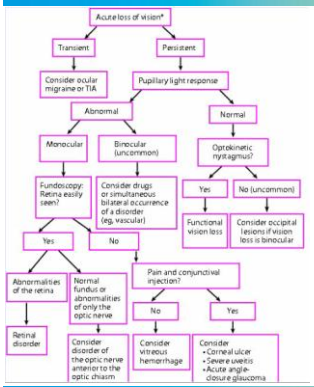


Differential diagnosis of optic neuritis

- Parainfectious optic neuritis
- Neuromyelitis optica Devic disease
  MAG or Aquaproin antibodies postive
  recurrent optic neuritis – optimal ttm wth Rituximab
- Multiple sclerosis
- Systemic disorders (as Sarcoidosis, Vasculitis, Lupus)
  - Systemic symptoms
  - Vasculitisscreen positive
- Leber's hereditary optic neuropathy
  - usually men/boys
  - Bilateral simultaneously or sequential, painless

Rebecca, 16 y old - previously healthy

- Bilateral deterioration of visual function within the last 3 hours
- Vision bilateral reduced to 40/100
- No improvement by glasses
- Pupils, ocular motricity, Optokinetic nystagmus normal
  Red color test and RAPD normal



References

Optic neuritis. Review
Lancet Neurol. 2014;13(1):83-99.
Toosy AT[1], Mason DF[2], Miller DH[3]

Diagnosing neuromyelitis optica.
Neuroimaging Clin N Am. 2013;23:279-91
Makhani N, Bigi S, Banwell B, Shroff M.

Parainfectious optic neuritis: manifestations in children vs adults.
J Neuroophthalmol. 2014;34):122-9
Rappoport D[1], Goldenberg-Cohen N, Luckman J, Leiba H.

Corticosteroids for treating optic neuritis.
Gal RL, Vedula SS, Beck R.
Cochrane Database Syst Rev. 2012 18;4:CD001430. doi:

Thank you for your attention