

TRANSVERSE MYELITIS

SUBTYPES

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Definition

- ✦ Intrinsic disorder of the spinal cord
- ✦ Temporal features
- ✦ Spatial features
- ✦ Etiology

Temporal and spatial patterns

- ✦ Peak reached at 10 days
- ✦ Damage in both longitudinal and horizontal planes
- ✦ All functions of SC are disturbed to various degrees

Semiology

- ✦ Prodromes
- ✦ Paresthesiae, paraparesis, backache, sphincter dysfunction
- ✦ Degree of paresis varies between slight weakness and total paralysis

Signs (cont.)

- ✦ Early, muscle tone is decreased (spinal shock), later spasticity develops
- ✦ DTR initially not obtainable or reduced, later they can brisk
- ✦ Babinski sign
- ✦ Trunkal sensory level

Etiology

- ✦ Parainfectious
- ✦ Postvaccinial
- ✦ Autoimmune
- ✦ MS
- ✦ Devic
- ✦ Paraneoplastic
- ✦ Vascular

Postinfectious & Postvaccinial Transverse Myelitis

- ✦ Prodrome
- ✦ Monophasic
- ✦ Autoimmune

- *Jeffery DR, Mandler RN, Davis LE. Arch Neurol 1993;50:532-535*

Etiology

Postinfectious

Rubella
Rubeola
Varicella
Influenza
EBV
Hepatitis A
Mycoplasma
Lyme
Syphilis

Postvaccinial

Rabies
MMR
Varicella
PPD
Influenza

CSF

- ✦ Pleocytosis (10-100 WBC)
- ✦ High protein
- ✦ Normal glucose

Infection

- ★ Viral: Echo, Coxsackie, HSV 1&2, Human herpesvirus 6, VZV, CMV, HIV, Hepatitis A, EBV, Mumps, Rubella
- ★ Bacteria: Mycoplasma pneumonia, Borrelia, Yersinia, Chlamydia, Rochalimaea, Mycobacteria
- ★ Parasites: Schistosoma, Cysticercosis, Toxocara, Toxoplasma gondii

Table III: Bacteria associated with ATM

<i>Infectious agent</i>	<i>Special characteristics</i>	<i>Diagnostics</i>	<i>Therapy</i>
<i>Borrelia</i> (Linssen et al. 1991)	Mostly radiculomyelitis, pure myelitis rare	Search for specific intrathecal AB synthesis. In exceptional cases first positive after 4 wk	High-dose penicillin G or third-generation cephalosporins for 21 days IV (e.g. cephtriaxon)
<i>Mycoplasma pneumoniae</i> (Heller et al. 1990, Mills and Schollfield 1992)	High fever, ESR accelerated, peripheral leukocytosis	AB in serum	Tetracyclines in combination with corticosteroids
<i>Yersinia enterocolitica</i> (Saebo et al. 1993)	Rare	AB in serum (ELISA)	Antibiotics
<i>Chlamydia psittaci</i> (Williams and Sunderland 1989)	Rare	AB in serum	Antibiotics
<i>Rochalimaea henselae</i> (Bogue et al. 1989)	In immunocompetent persons until now, only reports with simultaneous symptoms of cat-scratch disease	AB in serum (IF or EIA)	Gentamicin or cotrimoxazole, perhaps also other antibiotics
<i>Mycobacteria</i> (Bahemuka and Murungi 1989)	Only very seldom pure intrathecal forms; glucose concentration is reduced in CSF	MRI, intradermal test	Antituberculous chemotherapy, very rarely surgical therapy required

Table IV: Parasites causing ATM

<i>Infectious agent</i>	<i>Special characteristics</i>	<i>Diagnostics</i>	<i>Therapy</i>
<i>Schistosoma mansoni</i> and <i>Haematobium</i> (Haribhai et al. 1991, Nazer et al. 1993)	Sometimes intramedullary granulomas, also diffuse and necrotizing ATM	History of visiting infectious areas. If positive: serum AB (ELISA), MRI	Praziquantel, possibly corticosteroids in addition; laminectomy at CSF block or worsening while drug therapy is given
<i>Cysticercus cellulosae</i> (cysts from <i>Taenia solium</i>) (Kishore et al. 1991)	Eosinophile lymphocytes in serum and CSF	MRI; AB in serum and CSF (CF, ELISA); proof only by biopsy	Surgical removal of cysts, postoperative praziquantel
<i>Toxocara canis</i> (Linden and Berlitz 1994)	Rare, eosinophils in serum, IgE level raised	MRI; AB in serum (ELISA)	Thiabendazole or diethylcarbamazepine plus corticosteroids
<i>Toxoplasma gondii</i> (Wendefischer et al. 1993)	Rare, sometimes if patients are HIV positive, multifocal necrotizing course	MRI, CT, AB in serum. If HIV positive AB findings are difficult to interpret	Sulphonamides, pyrimethamin, spiramycin; plus corticosteroids

AB, antibody; ESR, erythrocyte sedimentation rate; ELISA, enzyme-linked immunosorbent assay; IF, immunofluorescence; EIA, enzyme immuno assay.

Table II: Viral causes of ATM*

<i>Virus</i>	<i>Special characteristics</i>	<i>Diagnostics</i>	<i>Therapy</i>
Varicella-zoster virus (Heller et al. 1990)	If infection is reactivated, mostly segmental paresis, rarely serious course. No invasion of spinal cord if associated with chickenpox. Consider VZV-ATM in immunosuppressed patients	PCR, AB search	If reactivation: acyclovir IV, if no improvement in course corticosteroids in addition
Herpes simplex virus 1 and 2 (Radhakrishnan et al. 1994)	Necrotizing myelitis	PCR, AB search	Acyclovir IV in combination with corticosteroids
Human herpesvirus 6 (Hill et al. 1994)	Rare	PCR, AB search	Symptomatic
B-virus/herpes simiae (Holmes et al. 1990)	Rare, survival only if therapy is started very early, until now only adult patients	Possible only if there was direct contact to monkeys. If history is positive try virus isolation from discharge from the wound, AB search in serum	Acyclovir IV
Epstein-Barr virus (Landgren et al. 1994, Junker et al. 1994 Tsutsumi et al. 1994)	Relatively good prognosis	PCR, AB search in serum and CSF (anti VCA-IgG and IgM, antiD, antiEBNA)	Symptomatic
Cytomegalovirus (Güngör et al. 1993)	Reported only in HIV-positive patients	PCR, search for pp65 in blood cells; conventional and shell vial culture	Ganciclovir IV, if resistance is suspected foscarnet
HIV (Geny et al. 1991)	Detailed diagnostics only if there is a positive history or risk factors	Diagnosis according to CDC criteria	Systemic therapy; recommendations change quickly
Coxsackievirus (Berlit 1988)	Rare	AB search in serum and CSF	Symptomatic
Echovirus (until now only subtypes 2, 5, 19, 25) (Barak and Schwartz 1988)	Rare, symptoms and signs can resemble polio	Culture from CSF, stool, throat swabs; AB search in serum and CSF	Symptomatic
Hepatitis A virus (Tyler et al. 1986)	Rare, reported only in association with typical general symptoms	Hepatitis serology	Symptomatic
Rubella virus (Bitzan 1987)	Very good prognosis, not always contemporary general symptoms	PCR from serum and CSF, AB search in serum	Symptomatic

Autoimmune-Rheumat.



SLE



MCD



PSS



SS

Laboratory

- ✦ ANA
- ✦ Double stranded-DNA Ab
- ✦ Soluble Nuclear Antigen Ab
- ✦ Anti-Ro
- ✦ AntiSS/A and SS/B
- ✦ RF
- ✦ CBC,APTT, Cardiolipin Ab
- ✦ CSF in 1st 24 hours(Low glu,high protein,pleocytosis)

Lupus myelitis

- ★ 17% of SLE-TM patients presents with TM as first SLE symptom.
- ★ Fever as prodrome
- ★ Pathology: Thrombotic complications due to cardiolipin antibody

Elsberg Syndrome

- ✦ Sacral radiculomyelitis
- ✦ Urinary retention
- ✦ Hypo/paresthesia –sacral dermatomes
- ✦ CSF pleocytosis without elevated protein
- ✦ Primary HSV type-2 infection
- ✦ Prognosis-good

Anterior Spinal Artery Syndrome

- ✦ Posterior cord function is preserved
- ✦ Acute onset of motor symptoms with dissociated sensory symptoms
- ✦ Etiology (60 cases)
 - ✦ Unknown (14)
 - ✦ Spinal angioma (10)
 - ✦ Postinfectious (9)
 - ✦ A. spinal artery occlusion (9)

MS and TM

- ★ TM is common initial presentation of MS
- ★ MS presenting as an isolated spinal cord or brainstem syndrome is 42% (*Miller et al.*)
- ★ Lack of relation to antecedent infection
- ★ Polyphasic course
- ★ Chronic recurrent
- ★ Progressive

Differentiation

- ★ 1. Symmetry of both sensory and motor impairments (>94% accuracy; *Scott et.al. Neurology 1998,50;2*)
- ★ 2. Oligoclonal bands in CSF

Complementary studies

- ✱ 3.MRI

- ✱ 4.Physiology

Prognosis

- ✦ Combination of severity of weakness and EMG had had 90.3% predicting power.
- ✦ Severity of weakness and denervation on EMG are most useful for predicting the outcome of TM at 6 months

Devic's Neuromyelitis Optica Diagnostic Criteria

- ★ C. Acute involvement of spinal cord and optic nerves, coincidental or separated
No cerebral, cerebellar or cortical features
- I. Normal brain MRI, enlarged, cavitated spinal cord
- L. No OCB-No CNS IgG-Incr CSF alb

Devic's NMO (cont'ed)

★ Pathology:

Necrosis, cavitation of spinal cord, thickened vessel walls
no inflammatory cells.

No plaques in brain, brainstem or cerebellum

Devic's NMO

Therapeutic Protocol

- ★ W&H, CXR, BP, UA, HbA-1C, Lytes, CBC, Stool guiac, PPD, LFT
- ★ Solumedrol 500 mg IV bid/2 hours x5 days
- ★ Prednisone 1 mg/kg/d qam for 2 months
- ★ Azathioprine 2 mg/kg/d at 3rd week
- ★ Very slowly convert prednisone to qod

Acute paraparesis with:
Sensory loss
Disturbances of defecation/micturation
Symmetric flaccid paresis

Immediately (day 1):
Spinal and cranial MRI with contrast medium to
exclude tumour, abscess, haematoma, spinal
vascular malformation, disc prolapse, MS

If negative:
Electrophysiology: F-waves,
NCV, VEP, BAEP, SEP, MEP

If positive:
Specific surgical therapy

Confirms lesion elsewhere:
Peripheral lesion: GBS – if unable
to walk >5 m treat with plasma
exchange or IVIG
Multifocal CNS lesion
– consider MS

Confirms spinal lesion:
Spinal tap: cell count, protein,
oligoclonal bands to exclude MS,
GBS; PCR of CSF: VZV, HSV,
EBV, rubella virus; if virus-PCR
not readily available IVSPT should
be started in combination with
acyclovir

If virus-PCR positive:
Start specific therapy
(compare text)

If virus-PCR negative:
Start IVSPT (20 mg/kg
prednisone) for 3 days

To delineate further the specific aetiology of ATM the
following examinations are recommended:
Serology (S=serum): VZV (CSF), HSV (CSF), EBV
(CSF, S), coxsackievirus (CSF, S), echovirus (CSF, S),
rubella virus (S), *Borrelia* (CSF, repeat!), poliomyelitis
(S), ANA (S), RF (S)
Culture for virus and bacteria from CSF, blood, stool,
throat swab

Modify therapy
if necessary

Only if other ATM causes are suspected the following
examinations should be carried out:
Serology (material is serum): HIV, B-virus, hepatitis A,
lues, *Mycoplasma pneumoniae*, *Yersinia enterocolitica*,
Chlamydia psittaci, *Rochalimaea henselae*,
Schistosoma, *Cysticercus cellulosae*, *Toxocara canis*,
Toxoplasma gondii, complete serology for collagen
diseases
PCR: cytomegalovirus
TB intradermal test

