Oculomasticatory Myorhythmia

Shirley H. Wray, M.D., Ph.D.
Professor of Neurology, Harvard Medical School
Director, Unit for Neurovisual Disorders
Massachusetts General Hospital
Oculomasticatory Myorhythmic: A Unique Movement Disorder Occurring in Whipple’s Disease

Oculomasticatory Myorythmia

1963 Van Bogaert et al.
1975 Knox et al.
1986 Jankovic J.
1986 Schwartz et al.
1987 Grotta et al.
1988 Hausser-Hauw et al.
1990 Adler and Galetta
1995 Simpson et al.
Oculomasticatory Myorhythmia

Unique pendular vergence oscillations
Smooth rather than saccadic
Peak velocities for various amplitudes typical of normal vergence movements
Disjunctive, continuous, unaffected by saccadic effort, visual stimuli, or sleep
PVOs are independently and uniquely generated within the vergence system

Schwartz et al. Ann Neurol 20: 19, 677
Pendular Vertical Oscillations

PVOs are truly pendular and devoid of any rapid “jerk” phase. They differ from other forms of pendular nystagmus because

- Oscillations in z-axis (anteroposterior) rather than the x or y-axis
- Have greater amplitudes (5-25 degrees)
- Slower frequencies (0.5 – 1.5 v 2-4 Hz)
- Absence of palatal movement
Pendular Vertical Oscillations

Distinguished from the nystagmus of Parinaud’s syndrome, which is

Episodic

Provoked by voluntary saccadic eye movements, especially attempted upgaze

Has a high-velocity saccadic component
Fig 3. Eye movement recordings indicate a smooth rhythmic oscillation of the eyes that approximates 0.8 Hz, ranges over 20 degrees, and requires 800 msec. Electromyogram (EMG) recordings of the masseter and genioglossus muscles are superimposed on the eye movement recording to show that discharges of muscle potentials are synchronous with the convergence phase of the oscillation.
SPONT ANEOUS EYE MOVEMENTS
eyes opened in the dark

OD VERT

OD HORIZ

OS HORIZ

10 deg

10 sec
EYE POSITION

R

L

A

B

1 sec

20°
Fig 2. Peak velocities of the ocular oscillations (open circles = convergence; closed circles = divergence) and refixations (squares) of Patient 1 are plotted against the amplitude of the eye movements. Slopes for normal vergence (40) and normal saccadic eye movements (3) are indicated by the solid lines. Both ocular oscillations and refixations approximate normal convergence and divergence eye movements.
Whipple’s Diagnosis
Whipple’s bacillus “Tropheryma whippelii”

Duodenal biopsy: PAS stain with diastase
Non-intestinal tissues: electron microscopy
Polymerase Chain Reaction:
  – Tissue, blood, and other bodily fluids
In situ hybridization fluorescent rRNA probe
Whipple’s Disease

CNS Involvement: 6-16% reported series

Primary CNS: < 5%
  - Progressive dementia
  - Myoclonus
  - Supranuclear ophthalmoplegia
  - Hypothalamic involvement
  - Obstruction of the aqueduct of Sylvius
The causative organism *Tropheryma whippelii* is seen within macrophages in the parenchyma on PAS (periodic acid-Schiff).
The causative organism *Tropheryma whippelii* is seen within macrophages in the parenchyma on silver stains.
PCR Detection of Microorganisms in Tissue
PCR Amplification of 16s rDNA

Bacterial 16s rRNA Gene

-834 bp

p8FPL

-904 bp

p515FPL

-475 bp

p91E

284 bp

PW3FE

p808R

p13B

PW2RB

BROAD RANGE PRIMERS

PRIMERS SPECIFIC FOR WHIPPLES'S DISEASE
PCR Detection of the Whipple’s Bacillus

Positive Control

100 bp

100 bp

Patient Negative + Cont.

mw 1 2 3 4 5 6 7 8
References


See case 932-1