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KAAU

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Diplopia for Investigation



Mr. Edi Muryanto Danjoso, 41 year old Indonesian male patient admitted through ER with history of **double vision** and **difficulty in talking** for 2 days. Not known to have any medical illness.

The problem started with **upper respiratory tract infection** (cough, runny nose) for 1 week after he came from al-hajj, he took Panadol and the symptoms subsided then in next days he started have a double vision and difficulty in talking (suddenly).





CNS: There is no loss of consciousness, no tremors, no seizures, no photophobia,

No numbness in the face, no eyelid dropping, no saliva dropping but there is **nasal regurgitation just for fluid**, no food collection in vestibule of mouth, no dizziness or vertigo, no difficulty in swallowing, no hoarseness in voice, no proximal or distal weakness in upper or lower limbs, he controlled his bowel and the urination, no excessive sweating or dryness,

No neck or back pain.



- **CVS:** no palpitation, no chest pain, no cyanosis,
- **RESPIRATORY:** no dyspnea, or orthopnea, or PND, no cough, no hemoptysis. (Just after the admission 2 days he developed epistaxis)
- **GIT:** no abdominal pain, no vomiting or hematemesis, no diarrhea or constipation.
- **SKELETAL:** no joints pain or swelling, no bone pain,
- **HEMATOLOGY:** no bruises, no bleeding from any orifices in the body
- **UROLOGY:** no Dysuria, no frequency, no hematuria, no oliguria



- This is his 1st admission in a hospital, not know to have any medical illness (HTN, IHD, asthma, DM) -ve past surgical history, never had blood transfusion, no allergy to any medication known.
- -ve family history for any medical diseases: No DM, no HTN, no IHD, no asthma. No similar attacks
- He is working as a driver for a family; he is married and has 2 children
- Not a smoker, he does not drink alcohol, no recent travel.

On examination



- On examination
- Patient looks well, conscious, oriented of time and place and person, he is right handed
- Memory intact, patient has **nasal speech**
- Vital signs :
B.p : 130/86 R.R : 24 pulse :72/m regular
temp. :36.6 O sat. :98% room air





CNS examination

Cranial:

- Olfactory:
- Optic: he has double vision (**ophthalmoparesis**) in all visual field, Pupil reaction minimal and **sluggish in light reflex**.
- 3, 4, 6 nerves: there is **weakness in eye movements** in all directions, there is **partial ptosis bilateral**
- Trigeminal: deep and superficial sensations and motor part are intact
- Facial N.: face is symmetrical, wrinkles are intact, no mouth deviation or saliva dropping, nasolabial fold is intact
- Acoustic N.: hearing is intact.
- 9. 10 nerves: he can cough, swallow, uvula in the center, gag reflex intact.
- Accessory N.: he can turn his head against resistance to both sides; also he can shrug his shoulder against resistance
- Hypoglossal N.: no vasculature or atrophy in the tongue, he can protrude it (central) and move it to both sides, and push it against his cheek against resistance.



Motor

- By inspection: no abnormal posture, no muscle atrophy, or pigmentation, no tremors, no scars (in both lower and upper limbs)
- Power 5/5 in upper and lower limbs bilaterally
- Tone normal bilaterally in upper and lower limbs
- Reflexes: brachioradialis, biceps, triceps reflexes: **absent**
Knee, ankle reflexes: **absent**.
plantar reflex is **equivocal**.





- **Cerebellar signs:**
No nodding of the head, **nasal speech** (as mentioned above),
There is **horizontal nystegmus**,
impaired left **nose to finger test**,
impaired left **disdiadokinesia**,
impaired left side **heel to shin**,
impaired left side **rebound**,
no trunkal ataxia.
- **Gait:** **ataxic wide base gait**, Romberg test –ve (not sensory ataxia)
- **Meningeal signs** are -ve
- **Spine** No spine tenderness on palpation



(Other systems examination, briefly)

- **CVS:** S1 + S2 + 0
- **RESPIRATORY:** bilateral equal air entry, vesicular breathing, no added sounds
- **GIT:** abdomen soft. Lax, no tenderness or masses (neither superficial nor deep)
- No lower limb edema





Deferential Diagnosis ??

DDx



- organophosphorus poisoning
- Acute neuropathy ... (GBS or GBS variants)
- Multiple sclerosis
- Alcoholic cerebellar degeneration
- Cerebellar vascular insult CVA
 - lateral medullary syndrome
 - vertebro-basilar thrombosis
- neuromuscular junction (Botulism , Myasthenia gravis)
- Paraneoplastic syndrome.





Investigation done for this patient



- Vitals checked every 4 hrs
- Peak flow rate checked every 4 hrs
- U & E (within normal references range)
- CBC (within normal reference range)
- HIV, HCV serology **-ve**
- Chest x-ray: **N**
- EEG (unremarkable)
- CT brain **-ve**, repeated with contrast: also **-ve**
- ESR: within normal reference range
- MRI: **-ve**
- LP *
- EMG *
- VEP *





LP (CSF)

- CSF :

glucose	2.9	mmol/l		N (2.2 - 3.9 mmol/l)
protein	0.50	g/l	↑	(<0.45 g/l)
gram stain		-ve		
culture		-ve		
wbc		3/cumm		N
Rbc		640/cumm		N (traumatic)
oligoclonal band		-ve		

(Cytological-protein dissociation)



Electrophysiology

NCS: motor nerve conduction study was **N**

SNAP: **absent** for median and ulnar nerves

EMG: no evidence for myopathic or neurogenic changes



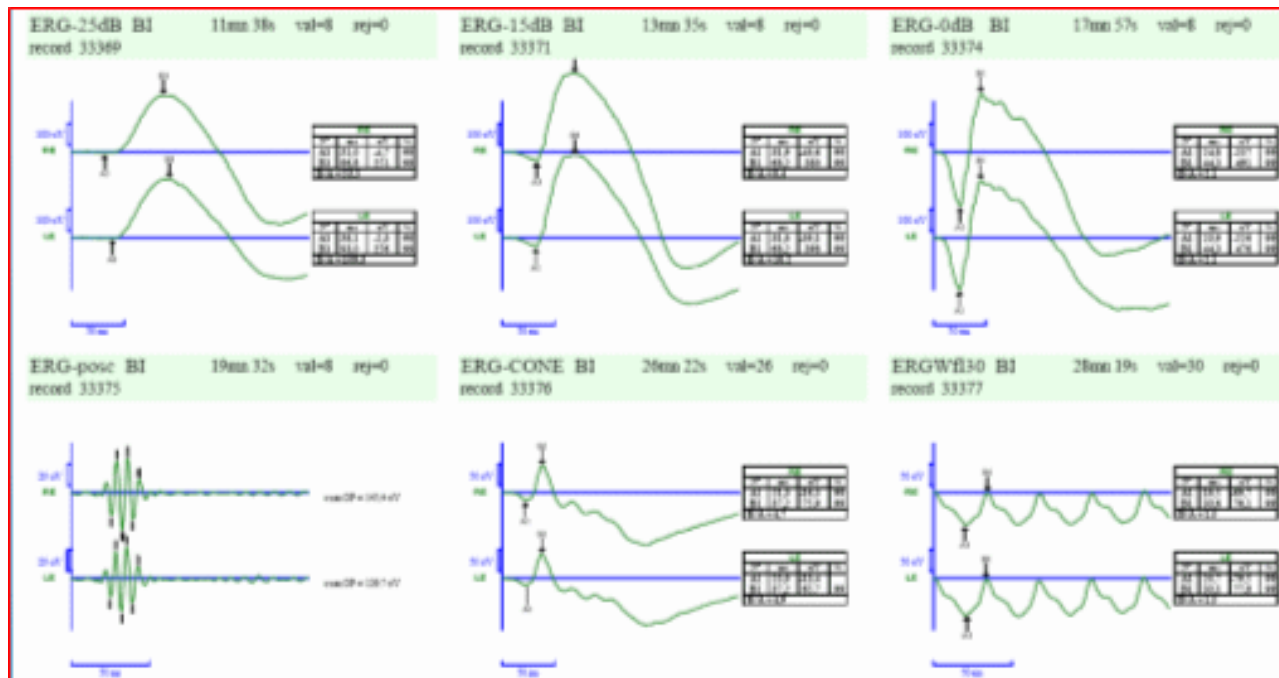


VEP

right eye : P100 latency of 123 ms

left eye : P100 latency of 116 ms

Normal (<100)



VEP

What is used for??

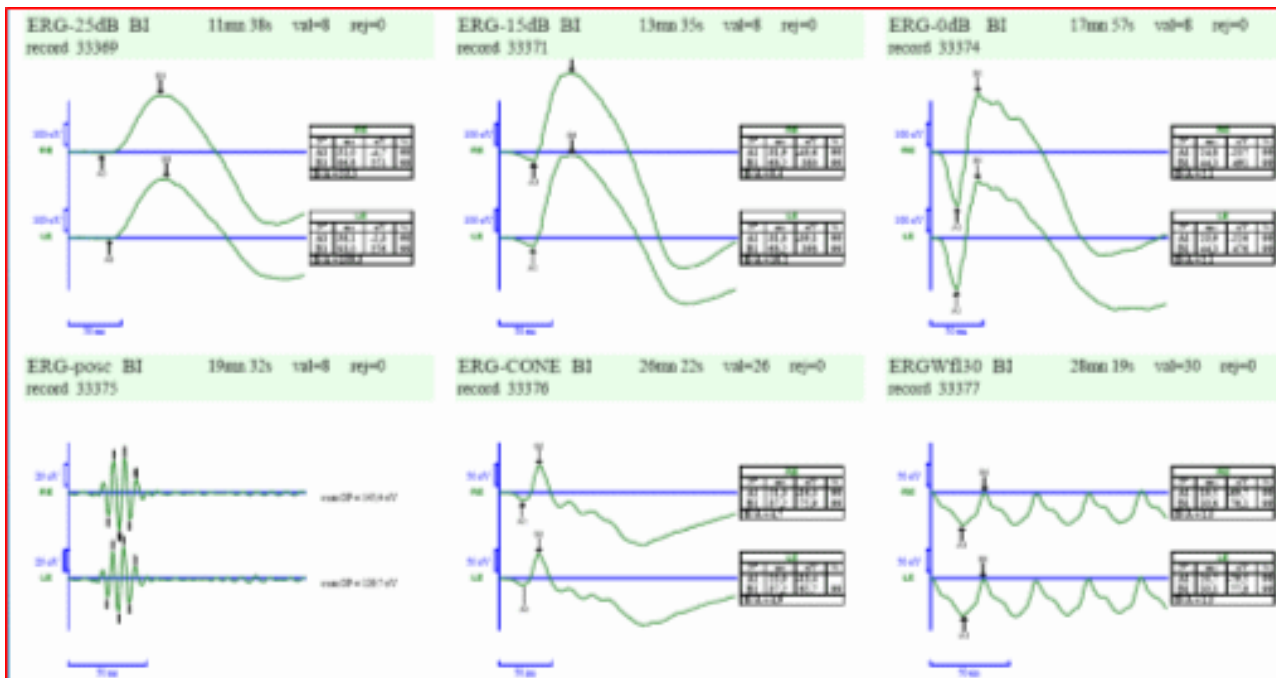
Any hints??



VEP : (detect of optic neuritis, delay signal)

right eye : P100 latency of 123 ms

left eye : P100 latency of 116 ms



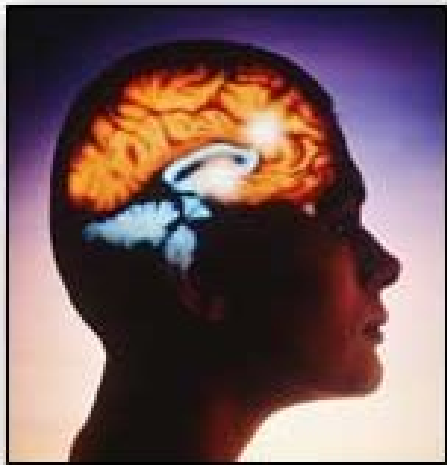
useful in detecting - **blindness** in patients that cannot communicate, such as babies or non-human animals

the diagnosis of **optic** - (**neuritis** (delay signal, MS

investigation of **basic** - **functions of visual perception**

Diagnosis !!!

Male, 41



Ophthalmoplasia

Ataxia

Areflexia

*preceded by viral infection

Slight High protein level in CSF
(cytological-protein dissociation)

Delay signal in VEP

**Miller fisher syndrome
variant of GB syndrome**



MFS

rapid overview





a variant of the Guillain-Barré syndrome

characterised by:

1. total external ophthalmoplegia
2. ataxia
3. loss of tendon reflexes

Reported in **male** patients between **38 and 65 years** of age. Complete recovery is usual.

Some authors have named the **Bickerstaff's syndrome** as a synonym for Miller Fisher syndrome, but that is a misnomer. Bickerstaff's syndrome is a brain stem encephalitis.

Additional Symptoms



- Unilateral or bilateral facial weakness
- Dysarthria
- Dysphagia
- Abnormal pupillary reactivity (as in our case)
- Extremity weakness

Trigger for Miller Fisher Syndrome: **viral infection**



Peripheral neuropathy

sensory nerve changes

histological examination → segmental demyelination of peripheral nerves

anti-GQ1b ganglioside antibody is detected in most patients

Detection by Latex Agglutination Assay or ELISA

GQ1b is highly enriched in the paranodal regions of human oculomotor nerves as compared with peripheral nerves.

Anti-GQ1b autoantibodies were further detected in cases of **Guillai-Barré syndrome with ophthalmoplegia** and in rare cases of **acute postinfectious ophthalmoplegia**



What is the prognosis?

The prognosis for most individuals is **good**.

recovery begins within **2 to 4 weeks** of the onset of symptoms, and may be almost complete within **6 months**. Some individuals are left with residual deficits.

Relapses may occur rarely (in less than 3%).



Treatments are similar to that for GBS

- Intravenous immunoglobulin (IVIg)
- Plasmapheresis (to remove auto-Ab)
- Supportive care



Break the Ice !!

MCQs





1. The average age of onset is

- A. 22.5
- B. 60
- C. 43.6
- D. 36

?



2. The following statement is true regarding MFS

- A. occurs more often in females
- B. reflexes are usually normal
- C. about two thirds of patients present with diplopia
- D. vertigo is a frequent complaint





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- A. occurs more often in females (M:F = 2:1)
- B. reflexes are usually normal
- C. about two thirds of patients present with diplopia
- D. vertigo is a frequent complaint (dizziness, not vertigo)



3. All of the following statements are true **except**

- A. the core syndrome consists of areflexia, ophthalmoplegia, and ataxia
- B. a viral infection often precedes the symptoms
- C. botulism can present in a similar fashion
- D. proximal muscle weakness is not seen in MFS





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- A. the core syndrome consists of areflexia, ophthalmoplegia, and ataxia
- B. a viral infection often precedes the symptoms
- C. botulism can present in a similar fashion
- D. proximal muscle weakness is not seen in MFS**

(Approximately 10% of cases may initially have mild proximal weakness)



5. All of the following are correct regarding MFS **except**

- A. the current belief is that the pathology is *central*
- B. campylobacter jejuni as well as antibodies to the ganglioside GQ1b are associated with MFS
- C. treatment is usually with plasmapheresis or intravenous immunoglobulins
- D. prognosis is good with recovery after a mean of 10 weeks

?



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- C. treatment is usually with plasmapheresis or intravenous immunoglobulins
- D. prognosis is good with recovery after a mean of 10 weeks

(Although brainstem encephalitis has been suggested to explain the pathology of MFS, the current belief is that the pathology is **peripheral**)



Done by **Dr.Wala'a Gholam**

KAAU, 2009
6th NNN Conference

Thank you for your time



www.medkaau.com/vb





معرض

بصمات أطباء عبر التاريخ

اثبت العلم والتاريخ أن لكل انسان "بصمة مميزة ثابتة لا تتكرر"
هناك من خلد بصمته عبر تاريخ الطب فكانت نقطة تحول كبيرة
في تاريخ الحياة البشرية.
ورغبة منا في تجديد الحماس و اثارة روح التحدي في اكتشاف الجديد ،
التعمق في بحور العلم ، وتحمل المسؤولية الملقاة على عاتقنا في تخليد
بصماتنا كأطباء مسلمين أقمنا هذا المعرض «

• الافتتاح :

الإثنين الموافق 16-3-2009

الساعة 12:00

بالمدخل الرئيسي لمستشفى جامعة الملك عبدالعزيز

• الأطباء والطلاب:

الثلاثاء 17-3-2009

• الصليبات والطلابات:

الأربعاء 18-3-2009

تنظيم:

رعاية:

