

ATAXIA, DIZZINESS AND NYSTAGMUS

Kamran Kabolizadeh

PGY 2

- ▶ 53 y.o WM with h/o HTN and hyperlipidemia transferred from an OSH on Jan 2008 to the MCV for further evaluation of vertigo for the past 3 days.
- ▶ Pt initially began having headaches for 2 wks prior to admission.

- ▶ Headaches associated with photophobia, nausea and gradually advanced to lightheadedness, ataxia, blurry vision and feelings of jumpy eyes.
- ▶ Denied slurred speech, dysphagia, diplopia, weakness and numbness.

- ▶ Social and family history not remarkable.
- ▶ Initially on physical exam:
MMSE=30/30, CN were intact with no nystagmus but purposeful saccadic eye movements on horizontal and vertical gaze with distractibility. Fundi were normal. DTR's were +2 b/l, downgoing babinski and normal parietal, cerebellar and sensorimotor exam. Gait was differed due to severe dizziness, vertigo and ataxia.

Medications: Diovan, pravastatin, roxicodone prn

Labs and imaging:

- ▶ BMP normal, CBC significant for mild elevation of WBC=11.9, UA negative, Cardiac enzymes negative.
- ▶ HCT: mild diffuse cerebral cortical atrophy.
- ▶ Normal sinus rhythm.
- ▶ TTE: EF=60-65%.
- ▶ MRI: negative for acute infarct.
- ▶ MRA: patent vessels.

- ▶ ENT recommendations: Assessment was complex migraine. Recommended valium prn dizziness and migraine.
- ▶ Neuro-ophthomology recommendations: Assessment was voluntary nystagmus. Recommendations were meclizine and compazine and discharge.

Second Admission:

- ▶ Pt readmitted to neurology again when he gained no improvement.
- ▶ On physical exam, found to have horizontal saccadic movement with bilateral nystagmus. Visual field and ophthalmic exam were normal.
- ▶ Pan CT of body was done to r/o neoplasm which was negative.

- ▶ LP showed protein=40, glucose=59, WBC=23/18, RBC=1/2 with negative cultures.
- ▶ Aseptic meningitis was suspected. Pt started on AB. ID was consulted who agreed with diagnosis but discontinued the AB.

- ▶ HIV, lymphocytic choriomeningitis antibody, TB were ordered by ID which were negative. Toxicology for arsenic, mercury and antimony were all neg. Paraneoplastic profile, C-ANCA, P-ANCA were neg.

- ▶ DDX: Brain stem stroke, neoplasm, vasculitis, toxic materials, labyrinths, aseptic meningitis, idiopathic opsoclonus, paraneoplastic opsoclonus, encephalitis secondary to HIV, TB and HSV.
- ▶ Diagnosis: Idiopathic opsoclonus was diagnosed.

Opsoclonus Myoclonus Syndrome

- ▶ The opsoclonus-myoclonus syndrome (OMS) is characterized by subacute onset of opsoclonus, a disorder of saccadic eye movements causing involuntary, chaotic saccades that occur in all directions.

Signs and Symptoms:

- ▶ opsoclonus (rapid, involuntary, multivectorial (horizontal and vertical), unpredictable, conjugate fast eye movements without intersaccadic [quick rotation of the eyes] intervals)
- ▶ myoclonus (brief, involuntary twitching of a muscle or a group of muscles)
- ▶ cerebellar ataxia, both truncal and appendicular
- ▶ dysphasia

Signs and Symptoms cont:

- ▶ mutism (a social anxiety disorder, in which a person who is normally capable of speech is unable to speak in given situations)
- ▶ lethargy
- ▶ irritability or malaise
- ▶ drooling
- ▶ strabismus (eyes are not properly aligned with each other)
- ▶ vomiting
- ▶ sleep disturbances
- ▶ About half of all OMS cases occur in association with neuroblastoma (a cancer of the sympathetic nervous system usually occurring in infants and children).

- ▶ OMS is a rare neurological disorder of unknown causes which appears to be the result of an autoimmune process involving the nervous system.
- ▶ Once paraneoplastic vs. idiopathic etiology for OMS was suspected, discovery of a possible underlying neoplasm such as neuroblastoma in children and small cell carcinoma or breast cancer in adults should be searched for.
- ▶ It is an extremely rare condition, affecting as few as 1 in 10,000,000 people per year

- ▶ Idiopathic OMS occurs in younger patients, the clinical evolution is more benign and the effect of immunotherapy seems more effective than in paraneoplastic OMS.
- ▶ In general, older pts had a slower improvement and were predisposed to remain with residual permanent neurological deficits, mainly gait ataxia.

- ▶ Evolution of the idiopathic OMS to coma and death is exceptional and usually reported in patients older than 60.
- ▶ Although idiopathic OMS in adults tends to follow a monophasic course, two cases were found to have chronic relapsing evolution highly responsive to IVIG.

Treatment:

- ▶ Paraneoplastic OMS has been empirically treated with a number of different therapies that caused a faster recovery.
 - ▶ Steroids and IVIG were reported to accelerate recovery.
- SCC is indicated to increase the chances of neurological recovery.

- ▶ Paraneoplastic OMS had a more severe clinical course, despite treatment with IVIG and corticosteroids.
- ▶ Treatment of the underlying tumor, usually SCLC is indicated to increase the chances of neurological recovery.

Prognosis:

- ▶ Idiopathic OMS occurs in younger patients and the clinical evolution is more benign and the effect of immunotherapy appears more effective than in paraneoplastic OMS.

Resources:

- ▶ The Opsoclonus Myoclonus Support Network recognizes the immediate need of opsoclonus myoclonus patients and their families to network with others affected by the syndrome, to have access to comprehensive information relating to the syndrome, and to be informed of latest research.
- ▶ <http://www.geocities.com/opso-myoclonus/>
- ▶ <http://www.omsusa.org/>

Board Questions

- ▶ Which of the following cancers were more commonly associated with paraneoplastic OMS?
 - a. Breast cancer
 - b. Small cell lung cancer
 - c. Non small cell lung cancer
 - d. Gastric adenocarcinoma
 - e. Lung cancer

- ▶ What residual deficit might stay with older patients after recovery?
 - a. Nystagmus
 - b. Vertigo
 - c. Gait ataxia
 - d. Confusion
 - e. Lightheadedness

- ▶ What's the best treatment for idiopathic OMS?
 - a. IVIG
 - b. Azathioprine
 - c. Methotrexate
 - d. Plasmapheresis
 - e. Treating underlying cancer