DANCING EYES

PRESENTED BY Dr. A. SHALINI - APOLLO CHILDREN'S

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Baby ‘one of the twins’

- 2 years 6 months/ Male / Chetpet
- Born late preterm by Elective LSCS
- Cried immediately after birth.
- Birth weight: 2.3 kgs.
- No neonatal problems.
- Normal growth and development.
In July 2014:

- Child presented to a local children hospital, Chennai with c/o vomiting of 10 days duration.
- Unable to walk without support since 1 week.
- Rolling movement in eyes 5 days, which worsened since 2 days.
History (cont)

- No h/o fever
- No h/o altered behaviour
- No H/o trauma/ drug intake
- No H/o recent vaccination
ON EXAMINATION (local hospital)

Documented to have

- Horizontal nystagmus+
- Titubation +
- Gait ataxia/ truncal ataxia+
- Fundus- normal.
Course: (local hospital)

CSF analysis - Normal
- MRI Brain and MRA - normal
Possibility of demyelination disorder and autoimmune disorder was considered.
- Was treated with IvIG
- No improvement.
- Patient got discharged

2/26/2015
Patient consulted multiple pediatricians/pediatric neurologist in Chennai and finally landed up with Apollo Children's Hospital in Sept 2014
ON EXAMINATION:

- Wt - 10.6 kg (below 3rd centile)
- Ht - 87 cm (below 3rd & 15th centile)
- PR - 124/min
- BP - 96/50 mmHg
- TEMP - 98.4 F

Eyes: multidirectional nystagmus present

SYSTEMIC EXAMINATION:

CNS: cranial nerves, motor system & sensory system - normal
Cerebellum: ataxic gait+, titubation +
# Differential Diagnosis of Abnormal Eye Movements

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<th>Pathology</th>
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<td>Rhythmic oscillations</td>
<td>Variable</td>
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<td><strong>Oposclocnus</strong></td>
<td>Non-rhythmic, chaotic conjugate movements</td>
<td>Neuroblastoma</td>
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<td><strong>Ocular flutter</strong></td>
<td>Intermittent bursts of rapid horizontal oscillations, during fixation</td>
<td>Cerebellar/ brainstem disease</td>
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<tr>
<td><strong>Ocular dysmetria</strong></td>
<td>Overshooting, undershooting or oscillations on refixation</td>
<td>Cerebellar disease</td>
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<td><strong>Ocular bobbing</strong></td>
<td>Intermittent, rapid downward movement</td>
<td>Pontine lesions</td>
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<td><strong>Periodic alternating gaze</strong></td>
<td>Cyclic conjugate lateral eye deviations, alternating from side to side</td>
<td>Posterior fossa</td>
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WHAT IS OPSOCLONUUS

- Opsoclonus consists of involuntary, non-rhythmic, chaotic, multidirectional saccades with horizontal, vertical and torsional components.
- Present during fixation, smooth pursuit, convergence and persists during sleep or eyelid closure.
- Also called as ‘dancing eyes’ syndrome.
- Often accompanied by myoclonic jerks of the limbs and trunk.
- Associated with Cerebellar ataxia, postural tremor, encephalopathy, behavioural disturbances.
ETIOLOGY OF OPSOCLONUS MYOCLONUS

- Paraneoplastic syndromes - Neuroblastoma.
- Parainfectious brainstem encephalitis - West nile, streptococcus, varicella zoster, lyme disease.
- Toxic/ metabolic states
- Idiopathic.
PATHOGENESIS

- Immune mediated, with a cross-reactive autoimmunity between neuroblastoma cells and the central nervous system.
- Several serum autoantibodies against neurons and cerebellar Purkinje cells have been identified in patients with OMS (specificity remains uncertain).
- Putative autoantigens reside in the cell surface or in the synapse and the antibodies cause transient neuronal dysfunction rather than permanent neuronal degeneration.
- Cell mediated immunity may also play a role.
INVESTIGATIONS

- CBC, liver, renal functions - normal

- USG abdomen - well defined, left suprarenal mass possibly pheochromocytoma/ neuroblastoma.

  (USG done previously was reported normal???)
INVESTIGATIONS

CT abdomen- well circumscribed, hypodense, homogenously enhancing mass, measuring 3.7*2.5*3.5 cms in the left adrenal region s/o neuroblastoma
MIBG scan

Rt ANTERIOR Lt
Lt POSTERIOR Rt
MIBG scan

Rt ANTERIOR Lt

Lt POSTERIOR Rt
MIBG scan

tracer concentration in upper pole of left kidney (compatible with neuroblastoma).
Pheochromocytoma/ neuroblastoma

- Urine VMA excretion by 24 hour urine - 1.2 (upto 13.6 mg/24 hours).
- 24 hour urine for nor-metanephrines - 142 µg/24 hours (50-111).
- 24 hour urine for metanephrines - 31 µg/24 hours (11-99).

- Spot urine HVA
- Chromogranin A - 136.2 (<98.1 ng/ml).
WORK UP FOR METASTASIS

- Nuclear Whole Body SPECT-CT-(Tc99m)
  soft tissue tracer concentration by the primary left adrenal tumour.
  No demonstrable abnormal skeletal lesion suggestive of metastasis.

Hematologist oncologist consulted
Surgery

- Child underwent laparoscopic removal of adrenal tumour in Oct 2014
HISTOPATHOLOGY

Biopsy:
stromal poor differentiating Neuroblastoma
PROGNOSTIC MARKERS

- Serum ferritin - 6.4 (reference range)
- Serum LDH - 259 U/L
- N-myc gene amplification (FISH) - negative

- Hemato-oncologist opinion: No chemotherapy
Interestingly, the prognosis for survival of neuroblastoma patients with opsoclonus is better than for those without opsoclonus.

Opsoclonus may represent an effective anti-tumour immunity that protects against tumour growth and dissemination.
Post-operative course - normal
Improvement in the opsoclonus on follow-up
Take home message

- Consider opsoclonus myoclonus as one of differentials in infantile and preschool kids with acute onset chaotic movement of eyes

- Exclude neuroblastoma in kids with opsoclonus myoclonus

- Surgery definitive treatment for neuroblastoma
Thank you.