DANCING EYES

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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.

COURSE IN OUTSIDE HOSPITAL

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 1week.
- 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

- Patient consulted multiple pediatricians/pediatric neurologist in chennai and finally landed up with Apollo Children's Hospital in Sept 2014

ON EXAMINATION;

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

Eyes: multidirectional nystagmus present

SYSTEMIC EXAMINATION:

CNS:cranial nerves,motor system & sensory system–normal

Cerebellum-ataxic gait+, titubation +

IFFERENTIAL DIAGNOSIS OF ABNORMAL EYE OVEMENTS

MOVEMENT	APPEARANCE	PATHOLOGY
NYSTAGMUS	Rhythmic oscillations	Variable
OPSOCLONUS	Non- rhythmic, chaotic conjugate movements	Neuroblastoma
OCULAR FLUTTER	Intermittent bursts of rapid horizontal oscillations, during fixation	Cerebellar/ brainstem disease
OCULAR DYSMETRIA	Overshooting, undershooting or oscillations on refixation	Cerebellar disease
OCULAR BOBBING	Intermittent, rapid downward movement	Pontine lesions
PERIODIC ALTERNATING GAZE	Cyclic conjugate lateral eye deviations, alternating from side to side	Posterior fossa

WHAT IS OPSOCLONUS

- 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

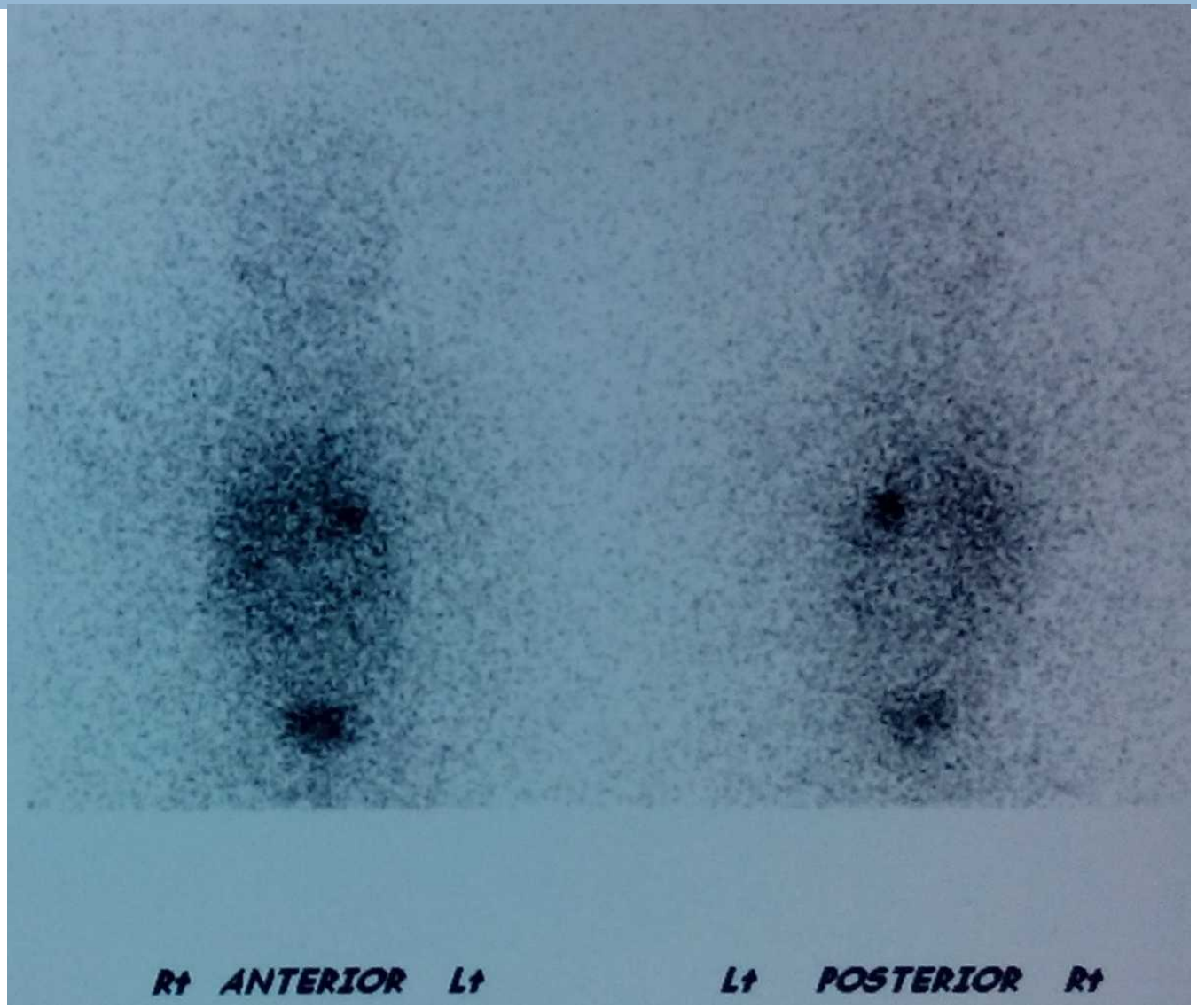
- USG abdomen- well defined, left suprarenal mass possibly pheochromocytoma/ neuroblastoma.

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

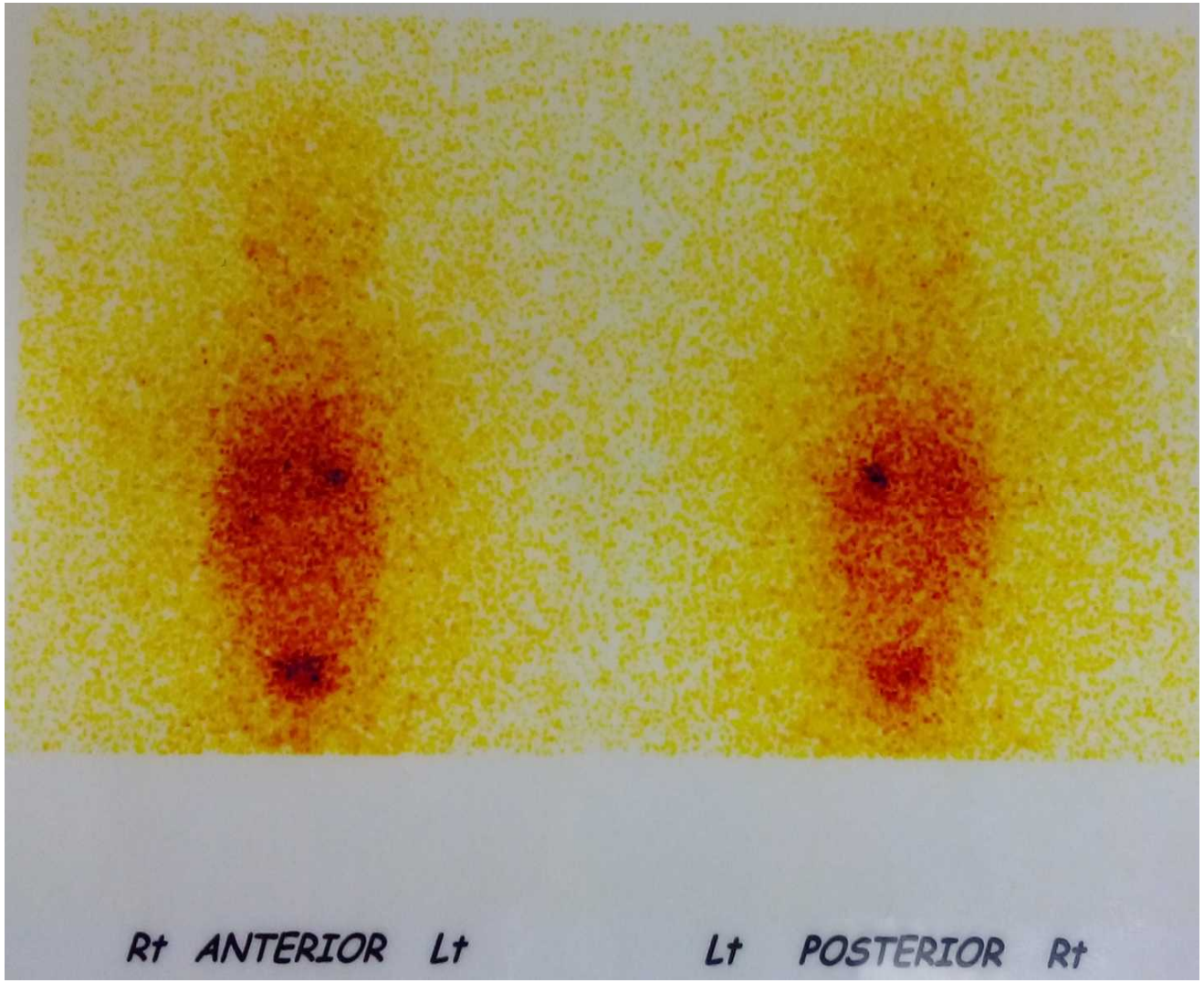
INVESTIGATIONS

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

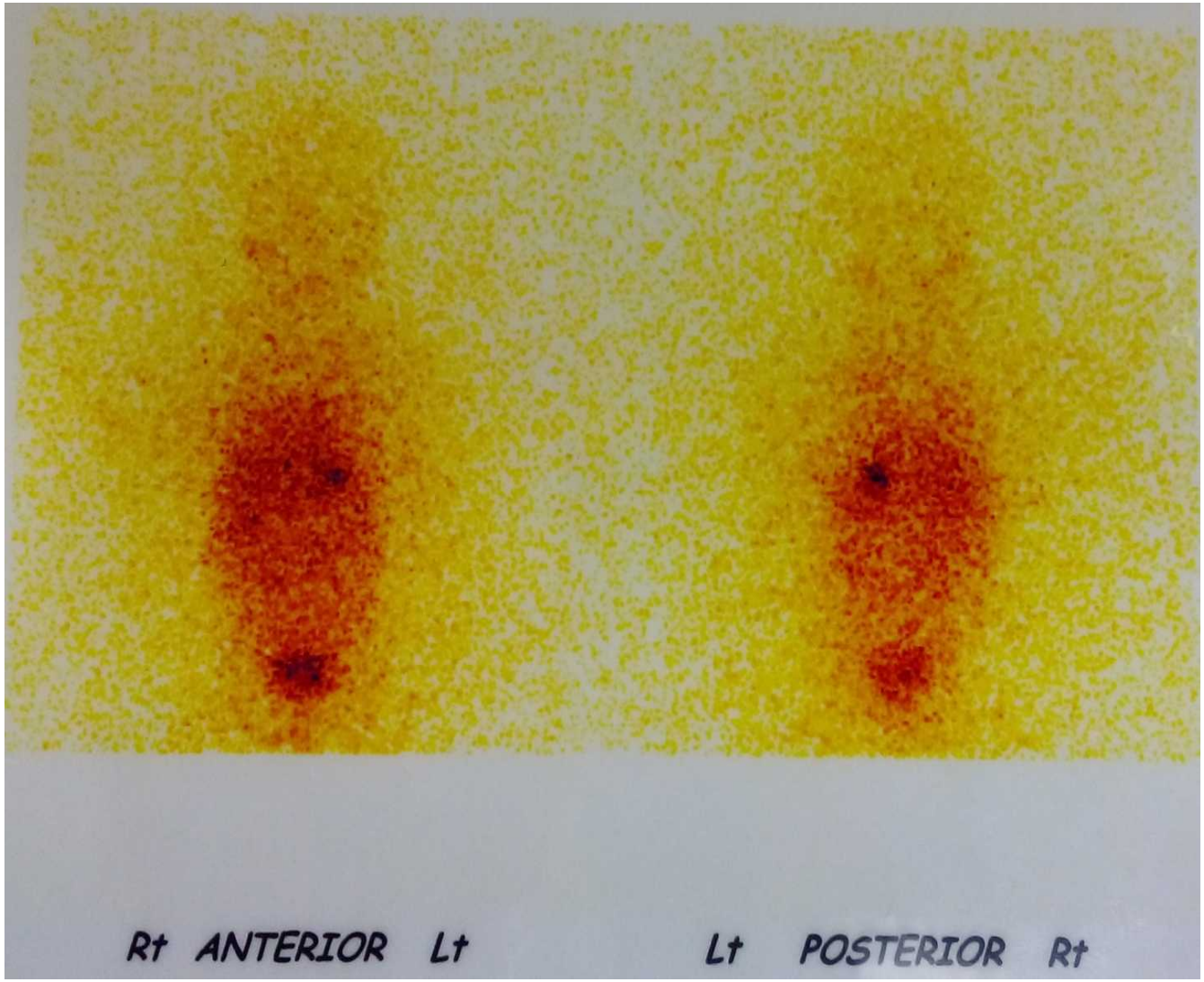


MIBG scan



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 13.6mg/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.

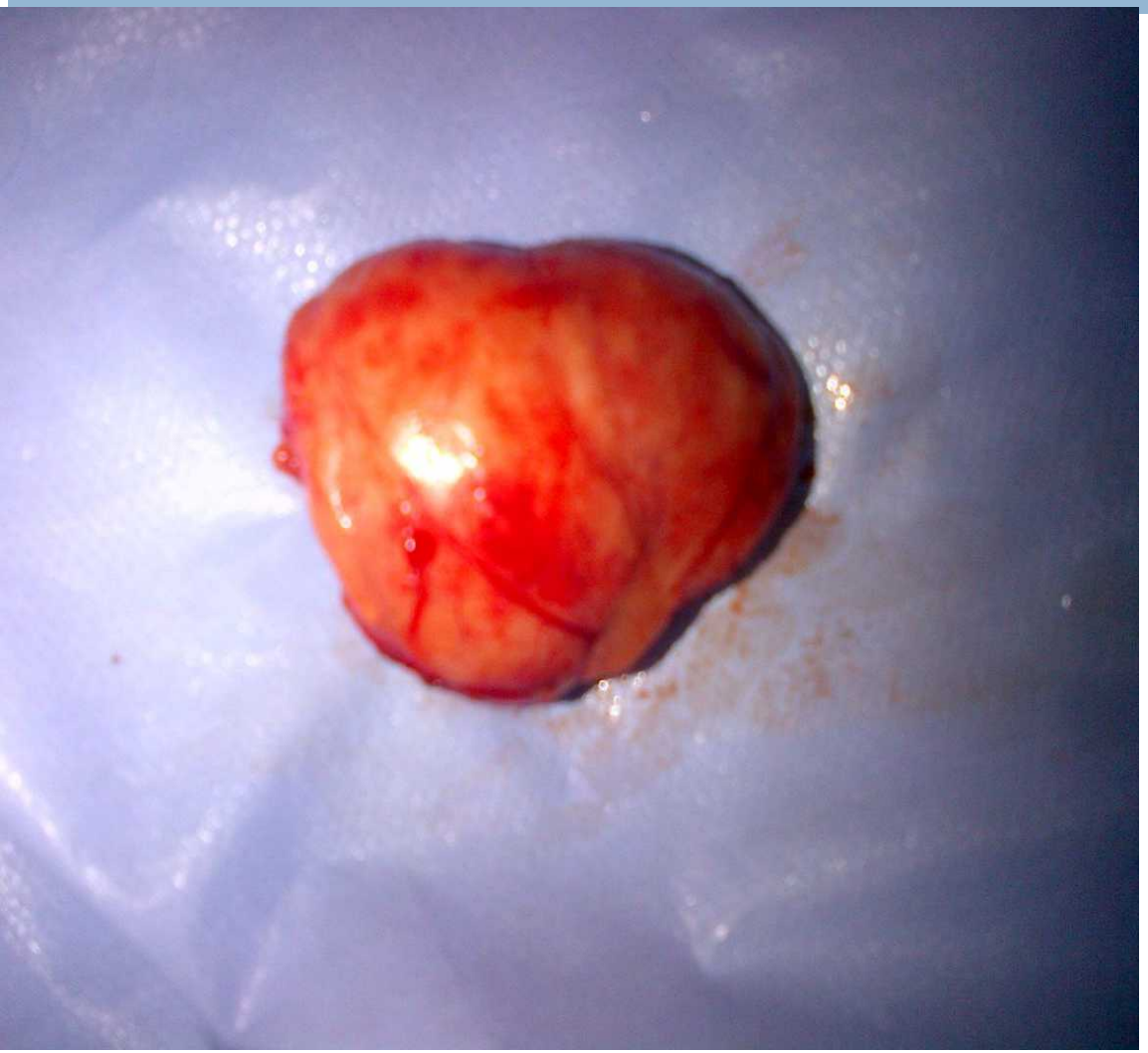
No demonstrable abnormal skeletal lesion suggestive of metastasis.

Hemato-oncologist consulted

SURGERY

- Child underwent laparascopic removal of adrenal tumour in oct 2014

HISTOPATHOLOGY



Biospy:

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

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TWIST IN THE TALE…..

- 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

COURSE

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 YO