AN UNUSUAL CAUSE OF LYMPHADENOPATHY IN INFANT

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Kanchi kamakoti CHILDS trust hospital

HISTORY…

- 5 month male infant, previously well
- swelling noticed over both sides of neck (right>left) x 3 months.
- Intermittent fever x 20 days
- Non consangunious, normal birth history

APRIL 2014

- Treated as suppurative lymphadenitis x 3weeks
- asymptomatic for a month
- gradual increase in size and reappearence of fever

Cervical lymph node biopsy

- granuloma with necrosis, epitheloid cells and giant cells
- S/O atypical mycobacterial infection or toxoplasma

KKCTH - MAY 2014

- well nourished child,no gross pallor
- Significant generalized lymphadenopathy- predominantly cervical-firm to hard consistency no tenderness
- liver palpable 2 cm below RCM
- No BCG Scar

Possibilities

- ?Disseminated TB
- ?Atypical mycobacterial infection
- ? Lymphoreticular malignancy

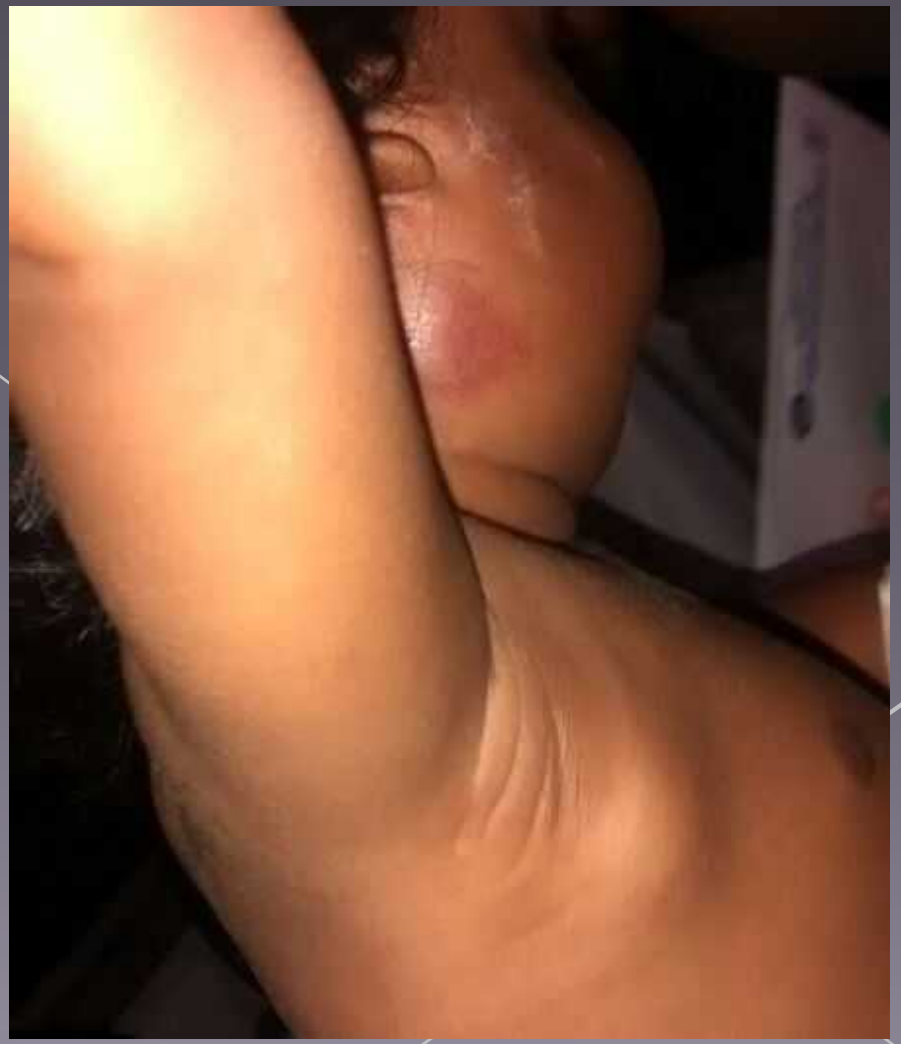
EVALUATION

- Leucocytosis , lymphopenia, anemia, thrombocytosis , no abnormal cells
- Toxoplasma IgM negative
- TB work up negative
- Immunoglobulin profile normal
- HIV Non Reactive
- Flow cytometry normal

WHAT NEXT....

- Awaiting second opinion on HPE of node biopsy, started on ATT

At review- JUNE 2014



SOME ANSWER....

- Lymph node biopsy HPE was reported as Langerhan Cell Histiocytoses

- NO skin rashes, otorrhea, irritability, fever, loss of appetite, diarrhea, weight loss, growth failure, polydipsia, polyuria, changes in activity level, behavioral and neurological changes.

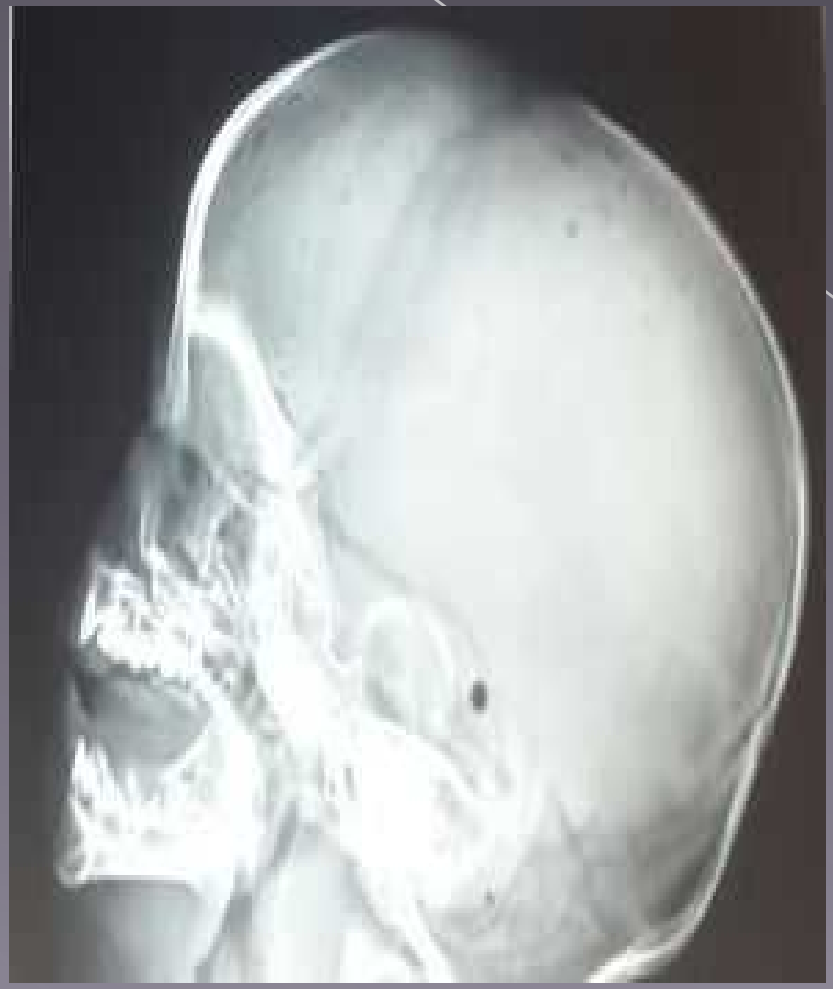
Further evaluation

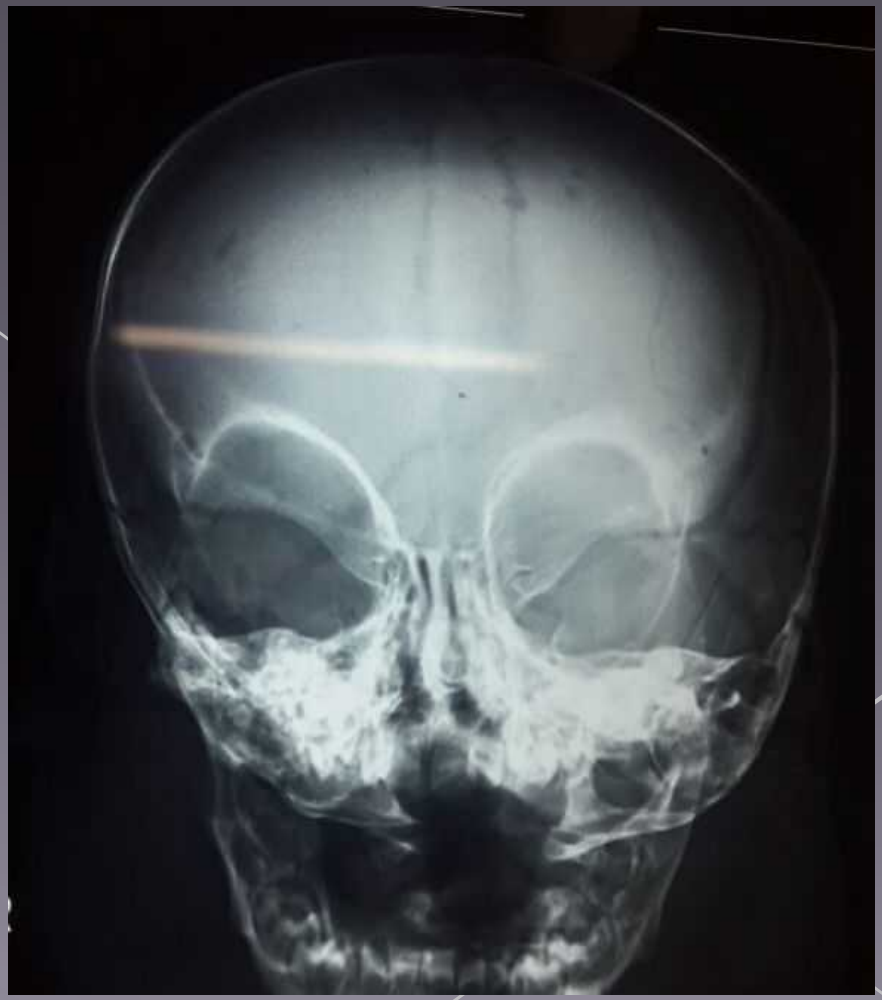
- Normal PT,PTT Ferritin
- Bone marrow – No infltration
- Hypoalbuminemia, normal bilirubin transaminases
- Normal RFT
- Urine osmolality - N
- USG abdomen- normal
- Normal TFT

- Right axillary node biopsy - suggestive of LCH

  Immunohistochemistry - strong, diffuse positive for S100, CD1a

Skull lesions…





FINAL DIAGNOSIS

MULTISYSTEM LCH

NO RISK ORGAN INVOLVEMENT

MANAGEMENT

- Started on chemotherapy (oral prednisolone and IV Vinblastine) as per Histiocyte society Evaluation and Treatment guidelines 2009

- Fever settled, cervical nodes decreased in size, clinical well being improved

LCH- Class I Histiocytosis

- Clonal proliferation of Langerhan cells, antigen presenting cells in skin (monocyte lineage).
- Electron microscopy- presence of bierbeck granule in the cytoplasm
- Contains langerin- involved in antigen presentation to T lymphocytes

Most common features…

- Skeletal involvement- 80 %
- Seborrheic dermatitis – 50 %
- Otitis media- 40%
- Lymphadenopathy- 33 %
- Hepatosplenomegaly- 20 %

Diagnosis…

- Histology- morphologic identification of the characteristic LCH cells.
- Positive staining of the lesional cells with CD1a and/or Langerin (CD207)- definitive diagnosis


LKS

Clinical stratification

Single System LCH (SS-LCH)	**One organ/system involved (uni- or multifocal):** • Bone: unifocal (single bone) or multifocal (>1 bone) • Skin • Lymph node (not the draining lymph node of another LCH lesion) • Lungs • Hypothalamic-pituitary / Central nervous system • Other (e.g. thyroid, thymus)
Multisystem LCH (MS-LCH)	**Two or more organs/systems involved** With or without involvement of "Risk Organs"

RISK ORGANS…

Hematopoietic involvement: (with or without bone marrow involvement*)	**At least 2 of the following:** • **anemia:** hemoglobin <10 g/dl, infants <9 g/dl (not due to other causes; e.g. iron deficiency) • **leukocytopenia:** leukocytes <4,0 x 10^9/l, • **thrombocytopenia:** platelets < 100 x 10^9/l
Spleen involvement:	• **enlargement** > 2 cm below costal margin in the midclavicular line
Liver involvement:	• **enlargement** > 3 cm below costal margin in the midclavicular line and/or • liver **dysfunction** (i.e. hypoproteinemia <55g/l, hypoalbuminemia <25g/l not due to other causes) and/or • **histopathological** diagnosis
Lung involvement:	• typical changes on **HR-CT** (low dose multi-detector CT if available) and/or • **histopathological / cytological** diagnosis

Single system LCH

- Usually spontaneous remission

- Currettage
- Low dose radiation
- Intralesional steroids

Systemic therapy indications..

- SS-LCH with “CNS-risk” lesions
- SS-LCH with multifocal bone lesions (MFB)
- SS-LCH with “special site” lesions

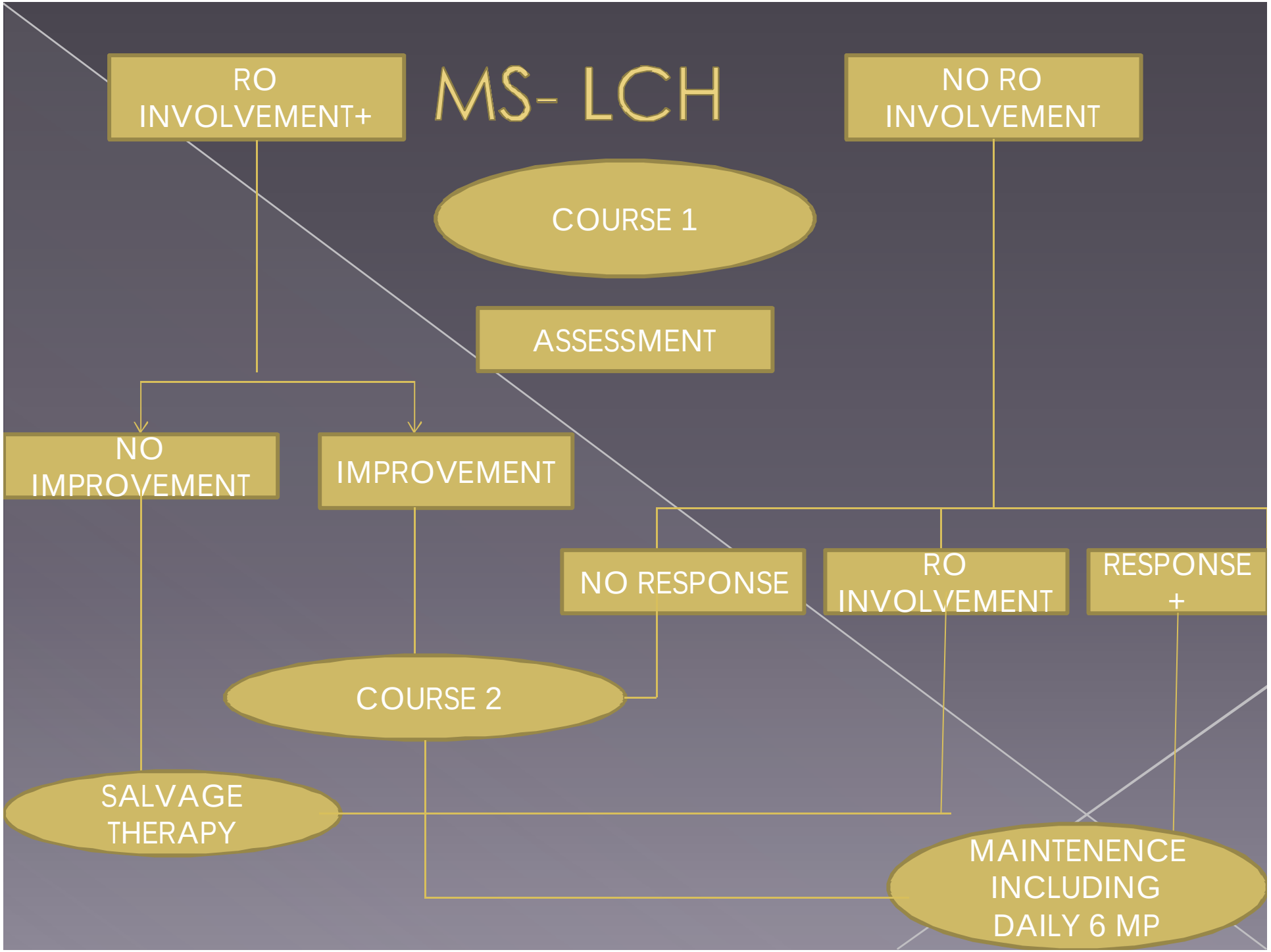
- MS-LCH with/without involvement of “risk organs

SS- LCH

6 WK-
WEEKLY VINBLASTINE +
DAILY PREDNISONE

6 WK-
WEEKLY VINBLASTINE+
WEEKLY PREDNISONE

12 months
3 WEEKLY VINBLASTINE+
3 WEEKLY PREDNISONE


MS- LCH
RO INVOLVEMENT+
NO RO INVOLVEMENT
COURSE 1
ASSESSMENT
NO IMPROVEMENT
IMPROVEMENT
NO RESPONSE
RO INVOLVEMENT
RESPONSE +
COURSE 2
SALVAGE THERAPY
MAINTENENCE INCLUDING DAILY 6 MP

SALVAGE THERAPY

- Not defined yet
- combined regimen of 2-chlorodeoxyadenosine and cytarabine (Ara-C)
- stem cell transplantation after reduced
- intensity conditioning regimen (RIC-SCT)

SUPPORTIVE THERAPY

- Pneumocystis prophylaxis
- Transfusions with filtered and irradiated blood products
- G-CSF

- management of complications – disease and therapy related

WHY THIS PRESENTATION…

- Lymphadenopathy – common feature of LCH
- LCH presenting solely as lymphadenopathy- relatively rare
- Histopathology may point to the diagnosis in case other investigations are non contributory.

THANK YOU…

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