

स्टरी की पती Instants जेवा कलाविसी साल आल अस्प्रसा Kalawati Saran Children's Hor

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युनिह Unit	270	5			सी, अ	तर, न, C.R. No	15958
HIH Name : Ayan	uh-	Ki	inne)				
आय Age :	2yr	Gm			लिंग	Sex Male	L
पता Address: <u>villa</u> 8431	30	Nas	war	<u>a</u>	Dist	shiv ha	s Bihar
भर्ती की तारीख : Date of Addmission निदान	1	5 [6]	13	_	खुद्टी ब Date of	की सारीख : (Discharge]	7/6/23
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LADY HARDINGE MEDICAL COLLEGE & SMT. SUCHETA KRIPLANI NEW DELHI DEPARTMENT OF RADIODIAGNOSIS

DEPARTMENT	TION NO: 28787
NAME: Ayansh AGE/SEX: 2y Y	Y/M REGISTRATION NO 2
REFERRED BY:U2C5 CT NO: 131/23	DATE: 9/1/23
CLINICAL DIAGNOSIS: K/C/Q Mutisystem	ie LCH. C/O respiratory distress not

CECT CHEST

SUBOPTIMAL SCAN DUE TO PATIENT MOTION. CT SCANNING OF THE CHEST WAS OBTAINED AFTER ADMINISTRATION OF INTRAVENOUS IODINATED CONTRAST. NO ADVERSE REACTIONS SEEN. STUDY REVEALS:

FINDINGS IN CHEST

- Bilateral lung fields appear normal.
- Trachea and major bronchi appear normal. Mediastinal vessels and cardiac chambers appear normal.
- No mediastinal lymphadenopathy seen.
- No pericardial or pleural effusion seen.
- Chest wall appear normal.
- In the visualised sections of the abdomen, hepatomegaly with linear hypoattenuating areas along the portal tracts ? portal triaditis. Advised USC correlation.

IMPRESSION: No significant abnormality in chest.

Please correlate clinically

Consultant

(wrapps

Dr. Shivani Senior Resident

Department of Pathology

G.B. Pant Institute of Post Graduate Medical Education and Research, New Delhi - 110002 (GIPMER)

Biopsy no: T12790/22

Year: 2022

Age: 2

Sex: Male

Name: AYANSH KUMAR

CR / OPD No: 28787

Referred By: DR NUPUR

Receipt Date: 24-12-2022

Specimen Received: T12790/22:Liver biopsy

Section examined show mainta nod lobular architecture. Six portal tracts identified show minimal chronic lymphocytic Inflammati ir No interface activity, Hepatocytes show mild and focally ballooning degeneration with non-zonal steatosis amount less than 10 percent. Sinusoids are dilated and infiltrated by histioc / -s with groove nuclei, few ill defined granulomas are also seen Masson Trichrome- No fibrosis Orcein- No copper associated p c din AFB- negative On IHC histiocytes cells are CDOE positive in sinusoids.

Impression: Feature are suggestive for histlic te storage disorders. T12790/22:Liver Kindly evaluate for Gaucher's distases.

Reported by:

Dr Puja Sakhuja/Dr RM(SR)

Verified by: Dr. AK

Date of Report: 09-01-2023

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6.3 Diagnostic Evaluation During Treatment and at Follow-up

The evaluation and the respective intervals and time points during therapy can vary depending on disease severity and treatment, and are therefore specified in the Roadmaps of each therapeutic Stratum (see Appendix A-IV).

The long-term follow-up evaluation scheme after end of systemic treatment is specified in Stratum VII (Section 14). The evaluation scheme is based on the assumption that patients have non-active disease (NAD) at the time being included in Stratum VII. In the case of complaints, signs and/or symptoms suggesting disease reactivation a basic evaluation as described in Section 6.2 has to be performed.

6.4 Definition of Organ Involvement

6.4.1 Risk organs

The definition of risk organs in the LCH-IV protocol is different from that of the previous study, since **lung** will **no longer** be considered **a risk organ**. The reason is the frequent association of pulmonary involvement with involvement of other risk organs, the low relative hazard ratio in a multivariate analysis, and last but not least, the very difficult and subjective evaluation of disease activity and therapy response in this organ.

A patient is considered to have risk organ involvement if at least one of the risk organs is involved. The current definition of involvement of the risk organs is presented in Table V.

Hematopoietic involvement: (with or without bone marrow involvement*)	At least 2 of the following: • anemia: hemoglobin <100 g/L (<10 g/dl), infants <90 g/L (<9.0 g/dl), not due to other causes e.g. iron deficiency • leukocytopenia: leukocytes <4,0 x10 ⁹ /l (4,000/µL) • thrombocytopenia: platelets <100 x10 ⁹ /l (100.000/µL)
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 dysfunction (i.e. hypoproteinemia <55 g/L, hypoalbuminemia <25 g/L, not due to other causes and/or histopathological findings of active disease

Table V: Definition of Risk Organ Involvement

Department of Pediatric Cardiac Sciences Sir Ganga Ram Hospital



DR. JAY R

DR. NEERAJ AGGARWAL

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DR. MRIDUL AGARWAL

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Ayomon 245) male

MD, (Peds), DM (Pediatric Carr Associate Consultant Pediatric Card E-mail jay.relan@gm DMC Regn No.

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A bone lesion with contiguous soft tissue involvement is continue

lesion.

ification of LCH

labre the	Definitions: trunk or multifocal):
Disease categories: Single System LCH (SS-LCH)	One organ/system involved (uni- or multifocal (>1 bone) • Bone unifocal (single bone) or multifocal (>1 bone) • Skin • Lymph node (not the draining lymph node of another LCH lesion) • Lungs • 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" (e.g. bematopoietic system, liver, spleen)

6.7 Stratification for the First-Line Therapy

Patients with indication for systemic therapy are stratified at diagnosis into two groups:

6.7.1 GROUP 1 - Multisystem LCH

 Two or more organs/systems involved, with or without involvement of "Risk Organs" (e.g. hematopoietic system, liver, or spleen)

6.7.2 GROUP 2 -Single-system LCH

- isolated "CNS-risk" lesion
- multifocal bone lesions (MFB) .

BIOCHEMISTRY SHEET

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¹⁸F-FDG WHOLF BODY PET-CT STUDY

Patient Name: AYANSH KUMAR		Age/Sex: 2Y/M
Study ID: FDG/23492/23	UHID:106391315	Date: 11.04.2023

Indication: Multisystem LCH, post Vinblastine (last-18.03.2023). PET-CT for disease

Procedure: PET-CT acquisition was done 60 minutes after injection of 10mCi¹⁰F-FDG by intravenous route, from the level of orbits to mid-thigh. CT was done for attenuation correction and anatomical localization.

PET-CT Findings:

Head and Neck: Increased tracer uptake noted in bilateral palatine tonsils with few subcentimetric bilateral cervical lymph nodes – infective Visualized paranasal sinuses, skull base, pharynx, larynx and thyroid do not show any abnormality on CT

Thorax: Few sub-centimetric bilateral axillary lymph nodes noted with preserved fatty hilum. Few paratracheal, prevascular, AP window, subcarinal and bilateral hilar lymph node noted, some of them showing calcifications, with no significant tracer uptake – like infective. Physiological FDG uptake is seen in the myocardium. Lungs, large airways, pleur heart, great vessels and other mediastinal structures appear normal on CT.

Abdomen-Pelvis: Hepatomegaly noted (CC span ~11cm) with mild FDG uptake all dilated intra-hepatic biliary radicles. Few sub-centimetric bilateral inguinal lymph no noted with preserved fatty hilum. Normal FDG distribution is noted in the liver, sp kidneys, gastrointestinal tract and urinary bladder. Biliary ducts, spleen, kidneys, stor adrenals, pancreas, retroperitoneum, bowel and urinary bladder appear normal on C ascites is noted.

Musculo-Skeletal System: FDG avid lytic lesions with soft tissue component n bilateral skull and facial bones. Opacification noted of right mastoid a Physiological FDG distribution is seen rest of the visualized axial and appendicular s

IMPRESSION:

- Metabolically active lytic lesions in bilateral skull and facial bones with right mastoid involvement-residual disease.
- No previous PET-CT available for comparison.

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Department Of Pathology All India Institute Of Medical Sciences Delbi

T= +91-11-26588500/26588700 Fax +91-11-26588500/26568700

Patient Name	Ayansh Kuman
F/H Name	Mukesh Kumar
Age/Sex	2 Y/Male
Clinic/Dept/Unit	Skin OPD/Unit 1
Reg Date:	13-12-2022

2252872 Acc. No. 106391315 Hosp Reg No UHID NO. Consultant Incharge Dr. N/A 22-12-2022 Reporting Date

Histopathology Report

Report Findings:

Received two specimens

1)Skin biopsy of chest papule shows unremarkable epidermis . At one end there is proliferation of langerhan cells abutting the epidermis with few eosinophils

Papillary dermis shows localised collection of langerhan cells histocytes.

2) Skin biopsy of back shows similar features with excess of langerhan cells. These cells are positive

for of CD1a, S100 and langerin

Overall features are suggestive of langerhan cell histocytosis

Reporting Incharge: Dr. Sudheer Arava

Reporting SR: Dr. Priya Jayakumar Dr. Priya Jayakumar Verify By











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KALAWATI SARAN CHILDREN'S HOSPITAL, NEW DELHI

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AT RANTA LCH (Multi-syster)

के बारे में अपनी मातू भाषा में समझा दिया गया है। यह रोग एक प्रकार का केंसर है। मेरा बच्चा जिस कैंसर में पीडित है, उसके इलाज और ठीक होने की संभावना के बारे में डॉक्टरों ने बता दिया है। इस विमारी के उपचार की अवधि लगभग

महीरी/Year है

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कीमोथेरेपी उपचार का मुख्य आधार है। इसके अन्य दुष्यमाय (side effects) हो सकते है। ये युष्यभाव स्थापी (permanent) या अस्थायी (temporary) ही मकते हैं आलो का झड़ना, भूख ज्वादा या कम लगना मुह में छाले, मुह में वदलाव होना, चिडचिडापन, पेट गर्द, कच्च, टागों में दर्द जादि जामतौर पर जस्याई (temporary) रूप से देखे जाने हैं कीमोधेरेपी के कारण अक्सर त्युट्रोपीनिया (सफेद Cell की कमी) हो जाता है। इसके कारण बुधार और संक्रमण (infection) होना आम बान है। मुख्यार होने पर तुरंत एटीबायोटिक (antibiotic) शुरू करना आवश्यक हैं कई बार संक्रमण (infection)) गंभीर और जानलेवा भी हो सकता है जरूरत घडने पर गंभीर समस्याओं के लिए इटेसिव केयर यूनिट (ICU) में जाने की आवश्यकता पड़ सकती है ICU में बैंड की उपलब्धि मांग और आपूर्ति पर निर्भर होती है कीमोधेरेपी कारण प्लेटलेट (platelets) की कमी हो जाती है जिसके कारण विभिन्न प्रकार की bleeding (खून बहना) हो सकती हैं कभी-कभी कीमोथेरेपी के कारण एलर्जिक रिएक्शन हो सकते हैं, जो गभीर रूप ले सकते हैं कदाचित (rarely), दवाईयों का असर दिमाग पर हो सकता है, जैसे दौर पहना, नसों में मुकसान, मस्तिष्क विकृति और मुद्धि में कमी, आदि कई दयाईयों के कारण हृदय (heart) को क्षति पहुंच सकती है जिगर या आंतडियों को नुकसान, मधुमेह, pancreas को क्षति, हडियो में कमजोरी, मैटाबॉलिक बदलाव, आदि दुष्यभाव हो सकते हैं ये दुष्यभाव इलाज के दौरान या समाप्ति के बाद देखें जा सकते हैं इलाज में कई प्रकार की प्रक्रियाएं आवश्यक होती है, जैसेकि बोनमेरी जांच और इटरथिकल (IT) कीमोथेरेपी, जिसके लिए एनसथिसिया (बेहोशी) जरूरी होता है एनसथिसिया (बेहोशी) आमतौर पर सुरक्षित होता है यदाकदा कई प्रकार की समस्याएं हो सकती है उदाहरण के तौर पर - सास का रुकना, दिल का दौरा, निमोनिया, आदि इलाज के दौरान खुन, प्लेटलेट (platelets) या पलाजमा plasma (खून का सफेद पानी) चढ़ाने की आवश्यकता पड़ सकती है कदाचित (rarely) इसके कारण एलर्जिक रिएक्शन हो सकते हैं और हिमेटाइट्स बी, सी, या HIV जैसे संक्रामक रोग भी हो सकते हैं कीमोथेरेपी के कारण प्रजनन (fertility) शक्ति पर दुष्प्रभाव हो सकता है इलाज के दौरान या उसके पश्चात, कैंसर के वापिस आने का जोखिम है

JAN SDHRT माता के हस्ताकर

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