BREAKING THE CODE OF A NODE

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&

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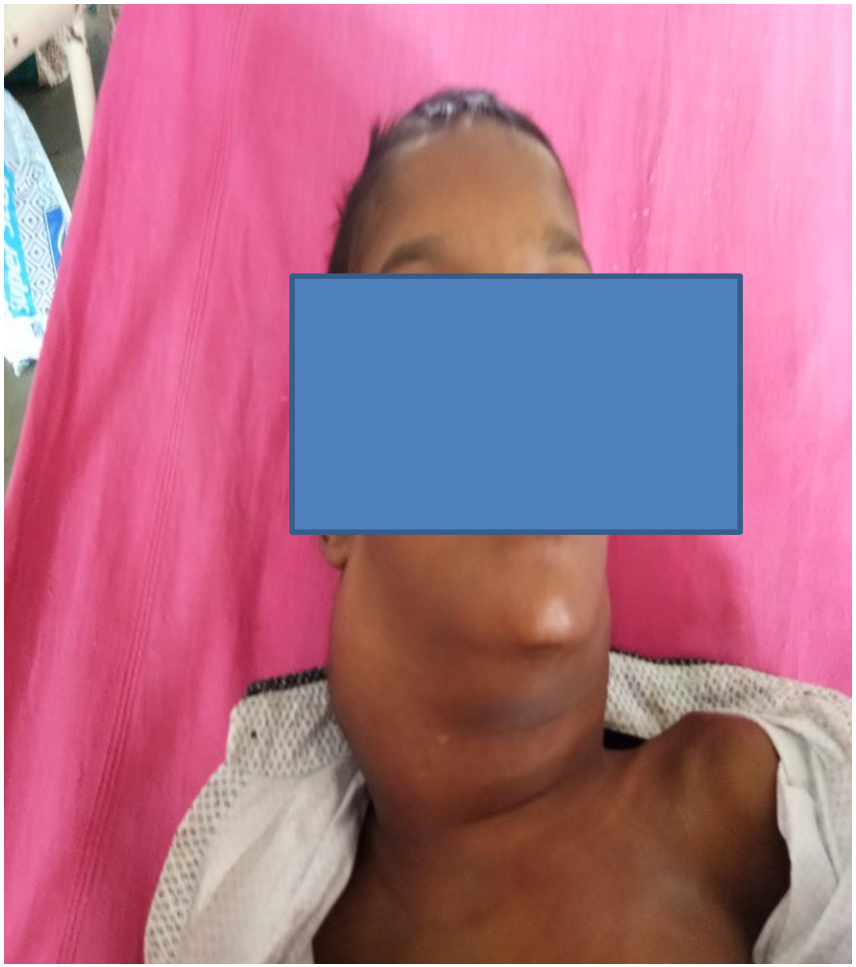
History of presenting illness

- 11 year old boy- second born of 3rd degree consanguineous marriage
- Brought by mother with complaints of swelling in the right side of the neck
- Progressive increase in size of swelling since October 2017.
- This was followed by similar complaints on the other side of the neck, which was not associated with pain.



- Right side 15cmx10cmx3cm,
- Left side 12cmx10cmx3cm, nodes,
- Firm to hard in consistency
- Neither tender
- Nor mobile.

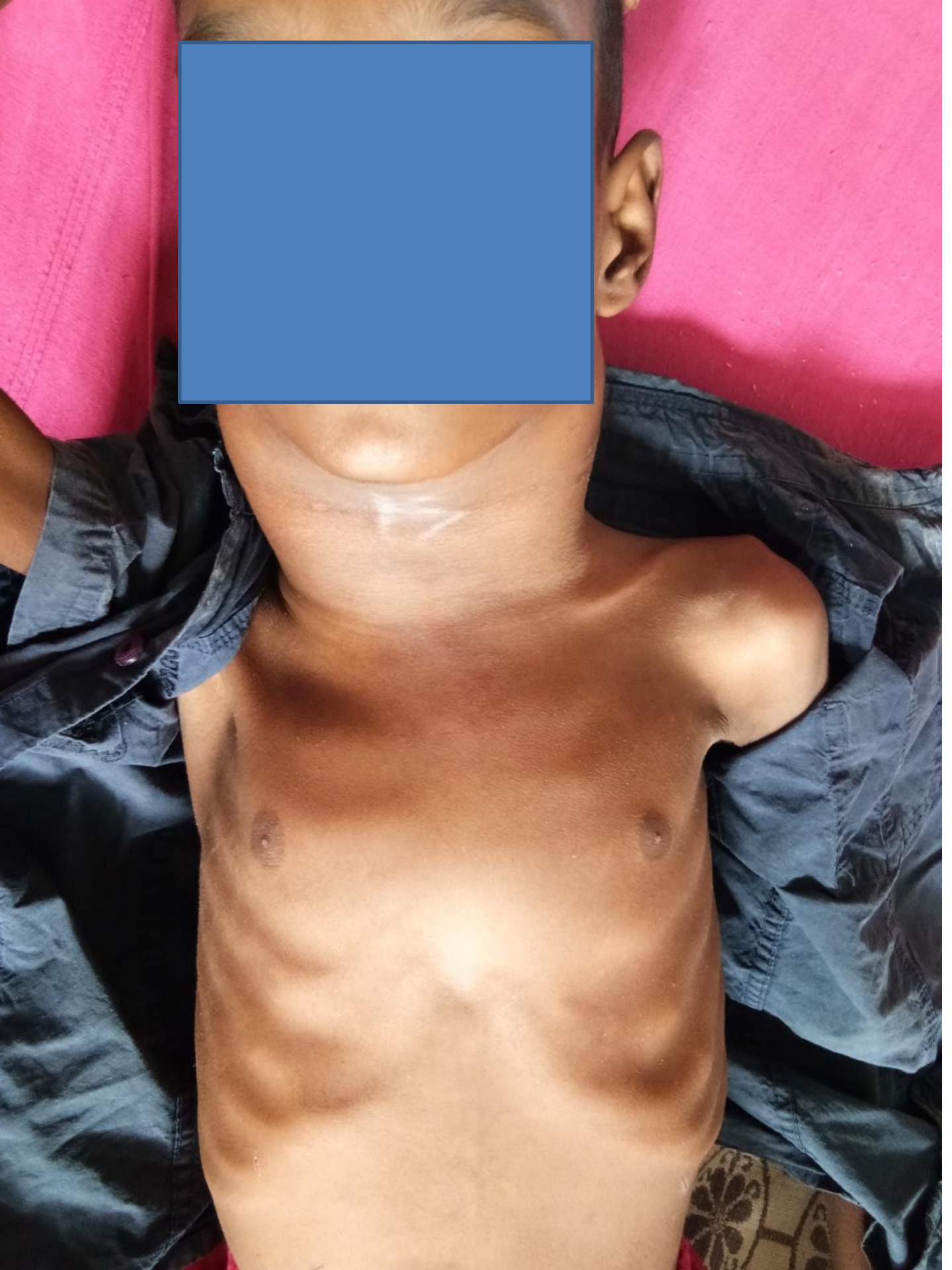
History of presenting illness

- h/o fever on and off for the past 3 months
- noisy breathing over the last three months.
- h/o easy fatigability
- h/o dysphagia
- h/o loss of weight 3kgs
- No h/o bone pain
- No h/o swelling of face
- No h/o skin lesions or ear discharge
- No h/o similar swellings elsewhere in the body

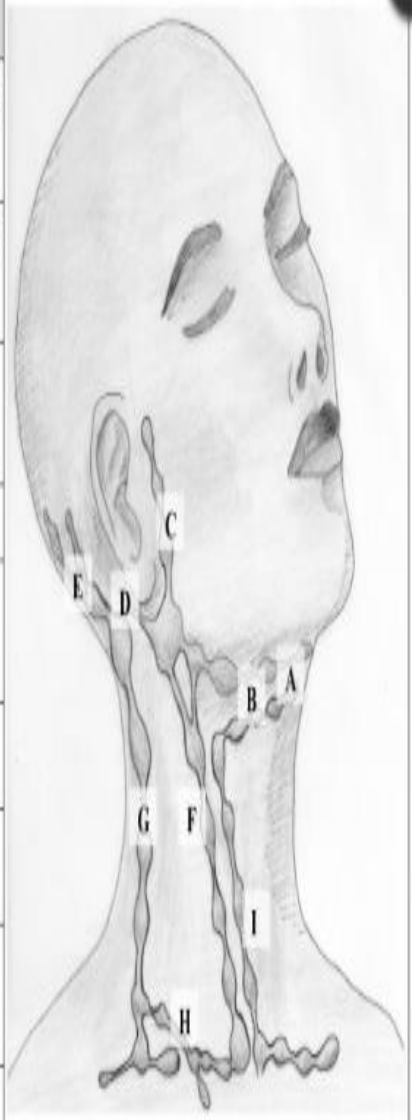
- **Past history:** no h/o previous admissions for similar complaints.
- **Ante-natal history:** uneventful
- **Natal and postnatal history:** child born by FTNVD, cried at birth. Birth weight of 2.5kg. No h/o NICU admission. Child had neonatal physiological hyperbilirubinemia

Other History

- **Immunization History:**
  - all vaccinations up to 10 yrs has been given.
- **Family History:**
  - no h/o similar illness in family members.
  - older sibling alive and healthy.
- **Contact History:**
  - no h/o contact with tuberculosis



	Region	Drainage areas	Predilection sites for:
A	Submental	Bottom lip, tip of tongue, floor of mouth, skin of cheeks	Infectious processes of the oral cavity, nose, maxillary sinus or the face
B	Submandibular	Tongue, submandibular gland, lips, mouth, conjunctiva	Infectious processes of the oral cavity, nose, maxillary sinus or the face
C	Pre-auricular	Eyelids, conjunctiva, temporal region, parotis, middle ear	Infectious processes of the ears, teeth, lid infections, conjunctivitis
D	Post-auricular	Auditory canal, auricle, scalp	Infectious processes
E	Suboccipital	Scalp, neck	Infectious processes, toxoplasmosis, rubella, lymphomas
F	Jugular	Tongue, tonsils, parotis, auricle	Pharyngitis, rubella, cancer metastases, lymphomas
G	Posterior cervical	Scalp and neck	Tuberculosis, melanomas, lymphomas
H	Supraclavicular	Lungs, esophagus, abdomen (thoracic duct)	Thoracic and retroperitoneal carcinomas, lymphomas, sarcoidosis, tuberculosis
I	Paratracheal	Trachea, thyroid gland	Lesion of the thyroid



Summary

Bilateral cervical adenopathy

Snoring

Fever

Weight loss

INVESTIGATIONS

- CBC:
- **TC-30,000/cu mm**
- DC-86/9/5
- **Hb-7g/dl**
- Pcv-21.3
- Platelets-6.07 lakhs
- **MCV-70.8fl**
- **MCH-23.3pg, MCHC- 32.9**, RDW-16.9, RBC COUNT -3 million, retic-2%

- **Peripheral smear**:
- Severe hypochromic anisopoikilocytosis, platelet in singles and good clumps, neutrophilic leucocytosis seen.

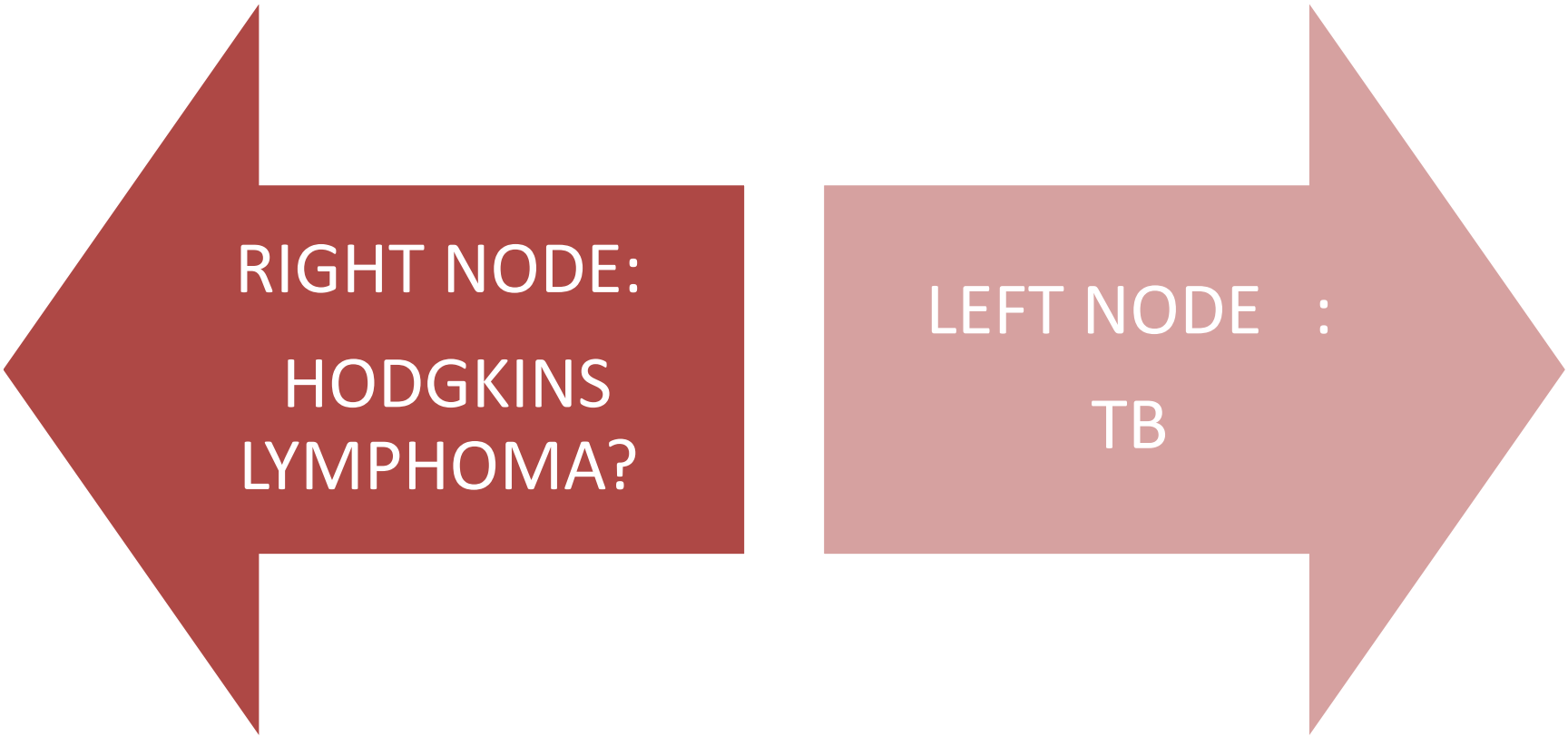
- Retroviral screening-negative,
- EBV SEROLOGY- negative,
- Chest x-ray normal study
- X-ray lateral neck- adenoid hypertrophy

- LEFT CERVICAL NODE EXCISIONAL BIOPSY
  - Granulomatous lymphadenitis with caseous necrosis
  - Suggestive of tuberculosis.

- Subsequent work-up for tuberculosis revealed no evidence of tuberculosis.
  - Mantoux Negative
  - CBNAAT Negative (GL/BAL)
  - Contact negative
  - CBNAAT from biopsy specimen : negative for TB

- Excisional biopsy Right cervical node

- Lymphoproliferative disorder suggestive of Hodgkin’s lymphoma


RIGHT NODE:
HODGKINS LYMPHOMA?
LEFT NODE :
TB

- Hence a repeat cervical node biopsy was done on 11/12/2017 showed evidence of atypical cellular infiltrate with surrounding fibrosis and acute inflammation .
- Possibilities :
  - Undifferentiated nasopharyngeal carcinoma
  - Germ cell tumor
  - Hodgkin's lymphoma

  And IHC was advised.

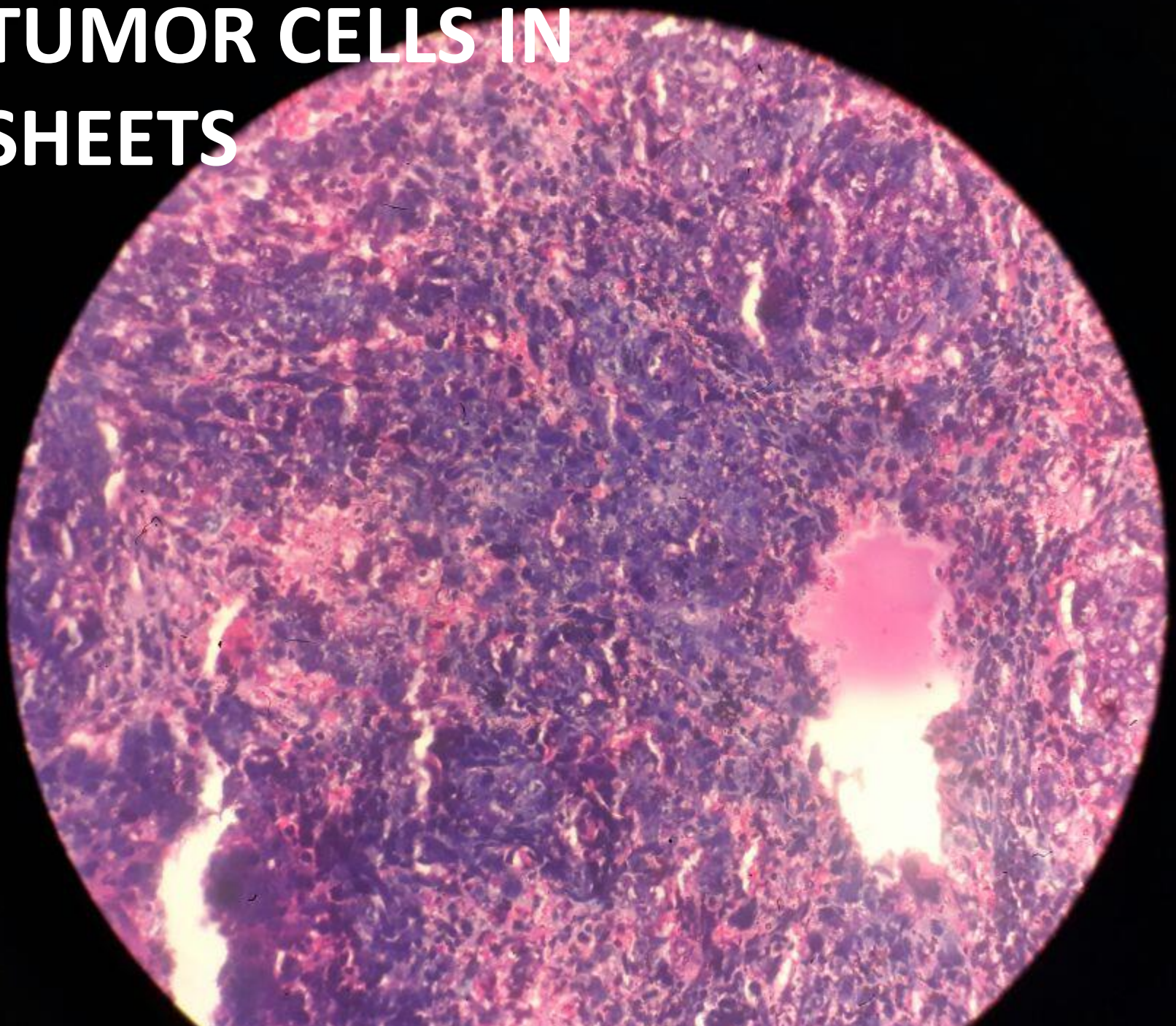
- **<u>IMMUNOHISTOCHEMISTRY:</u>**
- PAN CK: POSITIVE
- PLAP: NEGATIVE
- CD 30:NEGATIVE
- CD 15: NEGATIVE
- LCA: NEGATIVE IN TUMOUR CELLS AND POSTIVE IN LYMPHOID CELLS
- CK 5/6: NEGATIVE
- CK7: NEGATIVE
- CK19(A): NEGATIVE
- **IMPRESSION**: suggestive of **metastatic carcinoma**

- **ENT OPINION**: suggested CT neck and DNE was planned.
- **CT NECK**: bilateral level 2,3,4 and 5 cervical lymphadenopathy, enlarged retropharyngeal nodes 2.6cmx2.0cm with multiple necrotic areas. Impression: f/s/o ?lymphoma suggested HPE correlation
- **CT ABDOMEN**:normal
- **CT CHEST** : normal.
- UGI SCOPY: no suspicious lesion

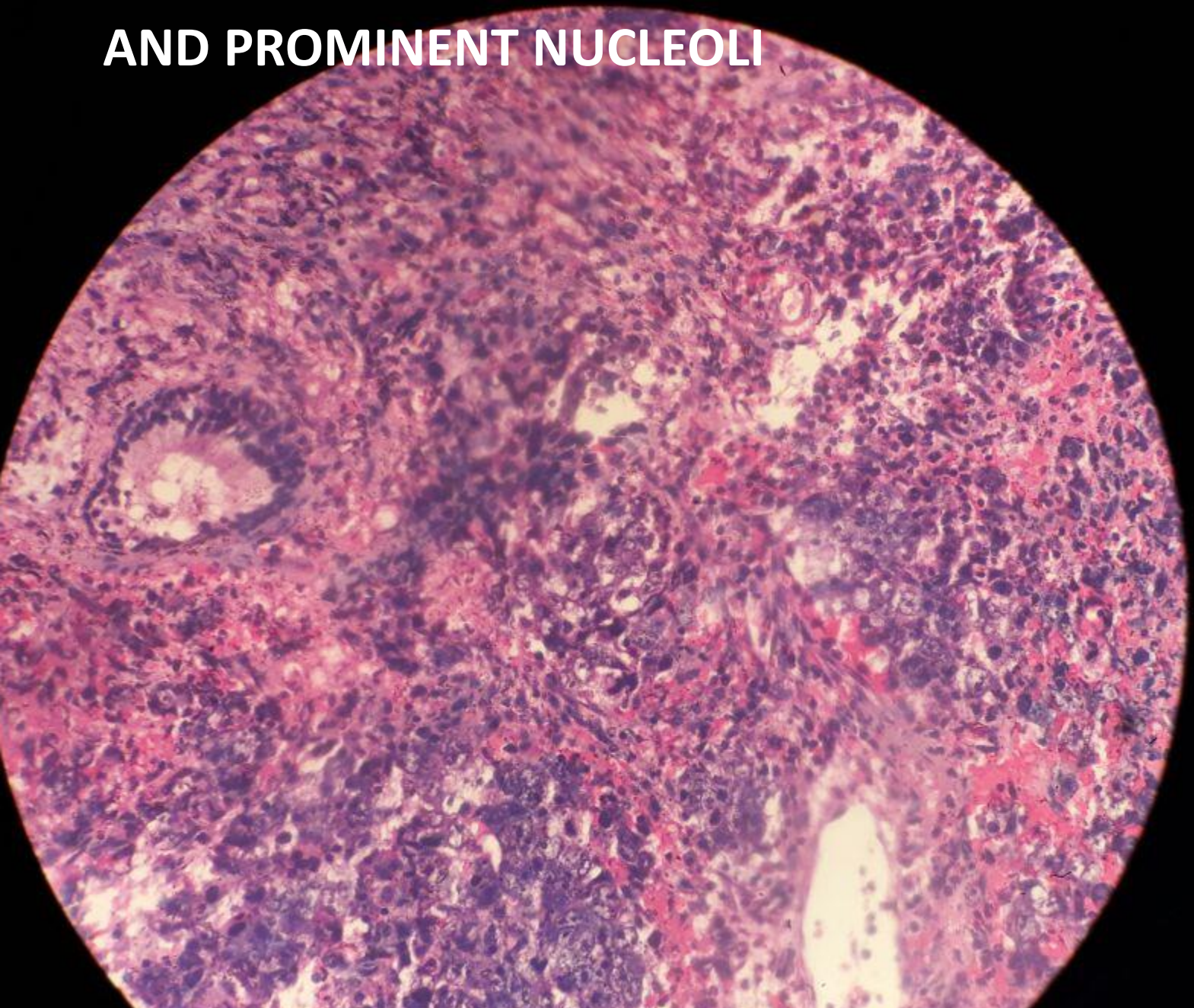
DIAGNOSTIC NASAL ENDOSCOPY

- Mucopurulent secretions in both nasal cavities
- Small friable mass visualised in the nasopharynx, biopsy taken from the same and sent for HPE.
- Grade three adenoid hypertrophy present.

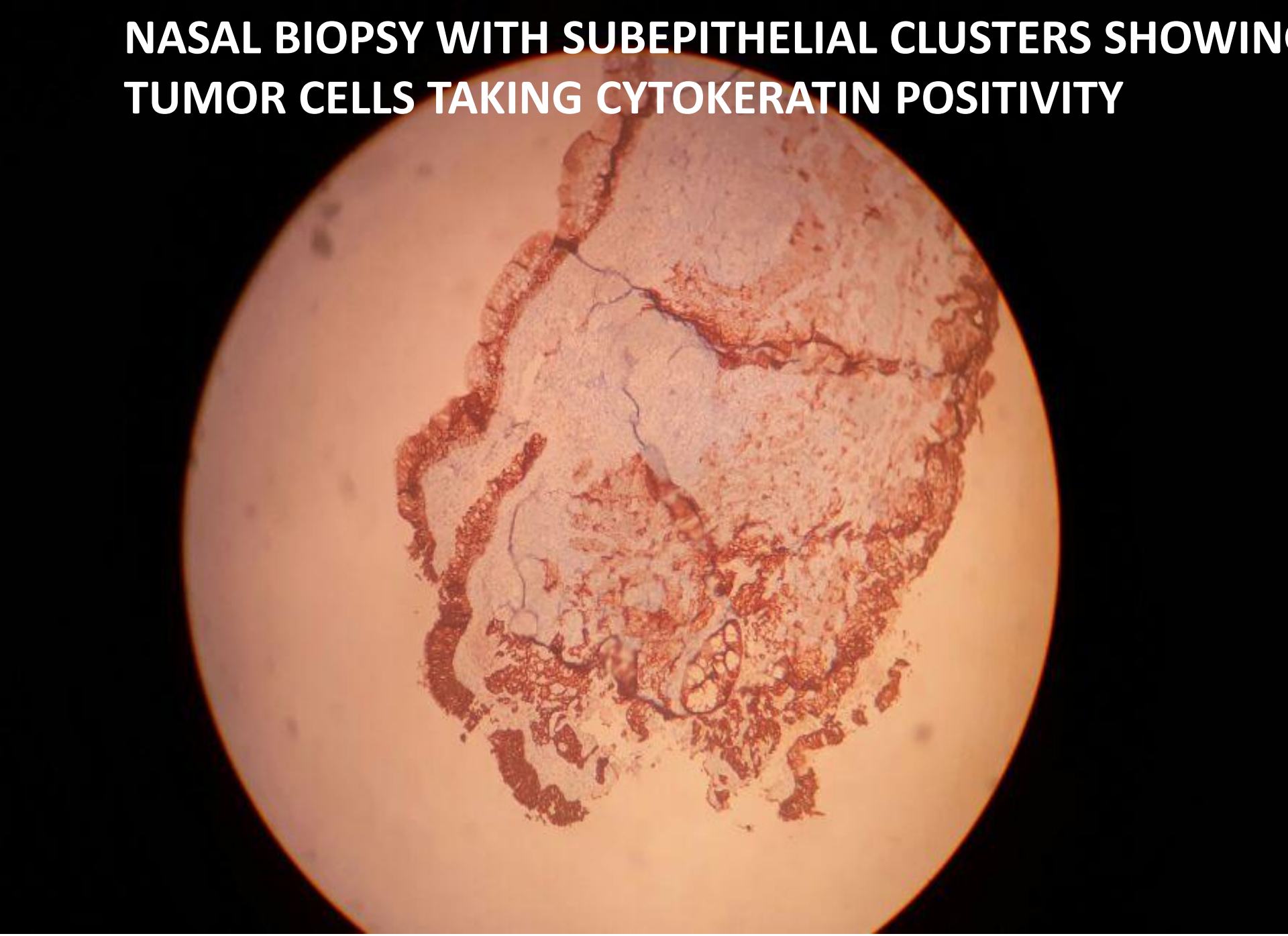
TUMOR CELLS IN SHEETS



TUMOR CELLS HAVING VESICULAR NUCLEI AND PROMINENT NUCLEOLI



NASAL BIOPSY WITH SUBEPITHELIAL CLUSTERS SHOWIN
TUMOR CELLS TAKING CYTOKERATIN POSITIVITY



- **Biopsy from DNE specimen:**

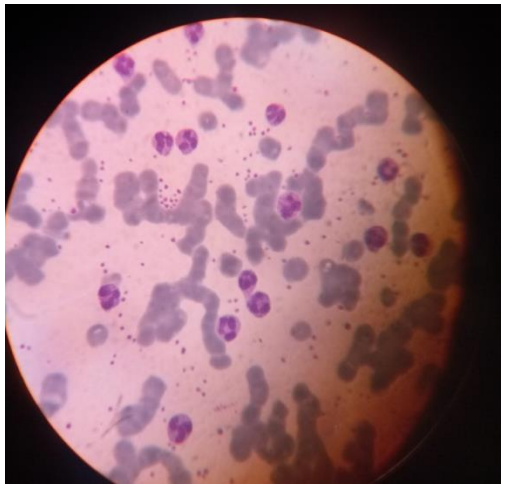
  suspicious atypical cell clusters which were subjected to IHC.
- **IHC with pan cytokeratin**: IHC section study shows strong and diffuse positivity in the tumor cells **favouring epithelial carcinoma**.
- Impression

  IHC from the primary was in favour of the diagnosis of **nasopharyngeal carcinoma.**

- During the course of stay child developed progressive increase in total counts.
- 28/1/18 **TC:68,200**, DC:**96**/03/1

- ONCOLOGIST OPINION
- : in view of deteriorating general condition child was started on hyperhydration and dexamethasone. Possibility of a lymphoblastic lymphoma was suggested.

- Peripheral smear on 29/1/18:
- RBC: NCNC+MCHC mild anisocytosis
- WBC: count markedly elevated with neutrophilic leukocytosis predominantly mature forms, no blasts seen.
- Platelets: thrombocytosis.
- Imp: leukemoid reaction,in view of clinical history with undifferentiated metastatic carcinoma of nasopharynx initially favours the diagnosis of **paraneoplastic leukemoid reaction.**



- leukemoid reaction,in view of clinical history with undifferentiated metastatic carcinoma of nasopharynx initially favours the diagnosis of **paraneoplastic leukemoid reaction.**

- Child was diagnosed to have undifferentiated metastatic nasopharyngeal carcinoma with paraneoplastic leukemoid reaction(after ruling out other causes of hyperleukocytosis in solid tumors).

- Child was shifted to department of medical oncology RGGGH and started on cisplatin and 5 fluorouracil and later radiation was planned

UNDIFFERENTIATED NASOPHARYNGEAL CARCINOMA WITH PARANEOPLASTIC LEUKEMOID REACTION

IHC IN NPC

- PAN CK- POSITIVE
- CK 5/6- POSITIVE(4/5)
- CK 8- POSITIVE(4/5)
- CK 13- POSITIVE(4/5)

PARANEOPLASTIC LEUKEMOID REACTION IN SOLID TUMORS

- Other causes of hyperleukocytosis-infections, new haematological malignancy, haemorrhage, corticosteroids and hematopoietic growth factors to be ruled out.
- Due to its rarity there are very limited studies regarding PLR.

- Largest study is a retrospective by Granger et al that observed 3770 adult oncology patients with hyperleucocytosis, among them 758 patients had solid tumors, only 77patients(10%) were found to have PLR. Of the 77 patients only 4 patients were found to have head and neck cancers.
- PLR is associated with heavy tumour burden and poor prognosis with an overall survival of 15 months.

THANK YOU

- METASTASIS SHOULD BE CONSIDERED AS A DD FOR CERVICAL ADENOPATHY
- PLR SHOULD BE CONSIDERED IN SOLID TUMORS WITH LARGE TUMOR BURDEN