# 北區重症討論會

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Admission time: 98/10/22
Age: 1-year-5-month-old
Growth:
BW: 9 kg = <3rd percentile
BH: 74.5 cm = ＜3rd percentile

## Present illness

- The 1-year-5-month-old boy was admitted due to aspiration pneumonia with recurrent wheezes and just discharged on 10/22.
- He was admitted again 10/22 due to shortness of breath after discharge.

- Trace back his history, he suffered from recurrent pneumonia since 4 m/o.
- Recurrent wheezy breath sound was usually noted and improved after bronchodilator inhalation.

## Admission history in CGMH

- 6/5-6/25
  - -HSV infection, CMV pneumonitis s/p anti-viral medication
  - -EBV infection (EB-VCAM positive)
  - -ITP
  - -GERD
- 7/24-7/30
  - -Aspiration pneumonia
  - -EBV reactivation
  - -parainfluenza III infection

- 8/4-8/15
  - -Aspiration pneumonia (MSSA)
  - -Recurrent wheezing favor hyperreactive airway
- 10/9-10/22
  - -Aspiration pnemonia
  - -recurrent wheezing

**In local hospital**

- Admitted 5 times due to recurrent wheezing with respiratory distress.

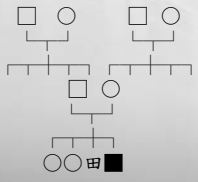
## Examinations done before

- Cadiac sonography :
  No positive findings
- Bronchoscopy:
  Suspicious lower airway infection
- Esophagography:
  frequent barium aspiration during swallowing→ r/o GERD

## Past History

1. Allergy: no known drug or food allergy
2. Operation history: no previous operation
3. Medication:
   steroid (10/10~) and bronchodilator

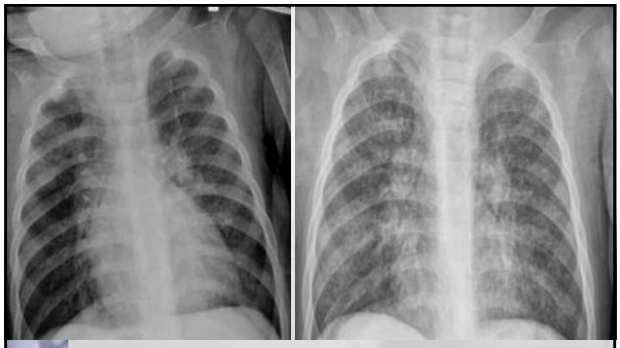
1. Birth history: G4P4A0, Gestational age: 39+4, NSD
2. Newborn screening: no abnormal finding
3. Development: delayed
4. Vaccination: as scheduled
5. Family history:
   His brother expired at 3y3m/o due to CMV pneumonitis, r/o VAHS (virus-associated hemophagocytic syndrome )



## Physical examination

T:38.6/℃ P:152/min R:30/min BP:103/60 mmHg
General Appearance: ill looking
Consciousness: clear, E4 V5 M6
HEENT: no positive findings
NECK:
supple, no lymphadenopathy
CHEST:
Suprasternal retraction
Breath sound:
bilateral wheezes, no stridor, no rhonchi

HEART:
Heart sound: regular heart beat, no murmur
ABDOMEN: no positive findings
EXTREMITIES:
Mild pitting edema over right lower extremities
Peripheral pulse: symmetric
NEUROLOGIC EXAMINATION: all negative findings



98/6/5

10/22

Reticulonodular infiltration in both lungs

| | | |
|---|---|---|
| WBC | 1000/uL | 19.5 |
| RBC | million/uL | 4.8 |
| Hemoglobin | g/dL | 8.7 |
| Hematocrit | % | 30.6 |
| MCV | fL | 63.8 |
| MCH | pg/Cell | 18.1 |
| MCHC | g/dL | 28.4 |
| RDW | % | 16.6 |
| Platelets | 1000/uL | 135 |
| Atypical-Lympho | % | 2 |
| Segment | % | 83 |
| Band | % | 8.5 |
| Lymphocyte | % | 3 |
| Monocyte | % | 3.5 |
| Eosinophil | % | 0 |
| Basophil | % | 0 |

| | | |
|---|---|---|
| BUN (B) | mg/dL | 11.5 |
| Creatinine(B) | mg/dL | 0.32 |
| AST (GOT) | U/L | 35 |
| ALT/GPT | U/L | 7 |
| Calcium | mg/dL | 7.9 |
| Inorganic P | mg/dL | 4.8 |
| Na | mEq/L | 134 |
| K | mEq/L | 4.5 |
| Cl | mEq/L | 99 |
| CRP | mg/L | 8.18 |
| Theophylline | ug/mL | < 2.0 |
| Mg | mEq/L | 2 |

## Impression

1. Suspect aspiration pneumonia with respiratory acidosis and impending respiratory failure
2. Recurrent wheezing favor hyperactive airway
3. Gastroesophageal reflux
4. Failure to thrive

- What will you do next?

## For r/o immunodediciency

- Check IgA, IgM, IgE, IgG (including subclass)

→slightly elevated IgE: 312 (IU/mL)

- T cell and B cell analysis

→No positive findings

## For rule out infection

- Vancomycin + Ceftazidime + Fluconazole
- 10/27 sputum culture (including fungus):

negative for bacterial, fungus

10/26 blood culture: negative

- Influenza A and B PCR: negative
- Sputum RSV, Chlamydia antigen: negative
- CMV-DNA/HSV-DNA: negative
- Throat virus culture: negative
- EBV DNA: positive→ add gancyclovir

- Due to impending respiratory failure he was transferred to PICU on 10/26.
- At PICU, after antibiotics, steroid, bronchodilator and high flow O2 support, symptoms improved →transfer to ward on 11/6
- Respiratory acidosis favor failure was noted again on 11/13 →transfer to PICU

| 檢驗項目 | 單位 | 981026 | 981103 | 981104 | 981113 |
|---|---|---|---|---|---|
| TEMP | ℃ | 37 | 37 | 37 | 37 |
| PH | | 7.314 | 7.502 | 7.529 | 7.234 |
| PCO2 | mmHG | 51.5 | 67.5 | 46 | 89.4 |
| PO2 | mmHG | 94.5 | 75.5 | 72.3 | 45.2 |
| HCO3 | mm/L | 25.6 | 51.7 | 37.5 | 36.9 |
| SBE | mm/L | -0.6 | 28.5 | 14.7 | 9.5 |
| SAT | % | 96.5 | 95.7 | 95.9 | 70.6 |
| Po2(A-a) | mmHg | 263.2 | 197.3 | 117.5 | 91.9 |
| FIO2 | % | 60 | 50 | 35 | 35 |

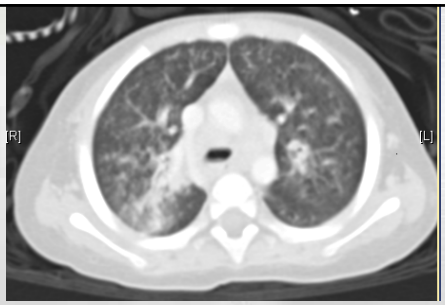
- 11/20-11/22 IVIG administration
- Keep ABx, steroid, bronchodilator.
- 12/4 transfer to ward
- Pneumothorax happened on 12/12 with respiratory distress

→transfer to PICU for pigtail insertion

| | |
|---|---|
| TEMP | 37 |
| PH | 7.202 |
| PCO2 | 96.2 |
| PO2 | 33.6 |
| HCO3 | 36.9 |
| SBE | 8.9 |
| SAT | 49.3 |
| Po2(A-a) | 205.4 |
| FIO2 | 50 |

**On Endotracheal tube with PCV**

- For recurrent wheezing with poor response to steroid and bronchodilator, we arranged chest HRCT.



Focal consolidation in RML, LLL, RUL (posterior segment), and multiple reticulo-nodular opacities (miliary nodules).

→Pulmonary tuberculosis with mediastinal lymphadenopathy is first consider. Lymphoproliferative disease should also be considered.

→3 set TB smear and culture were all negative

- What will you do next?

- Lung Biopsy was arranged:

  Sections showed lung tissue with diffuse interstital infiltration of small to large lymphoid cells with nuclear atypia.
- Gene rearrangement showed Presence of monoclonal B-cells in the lung biopsy→lymphoproliferative disease is more likely.

## Lymphoproliferative disorder

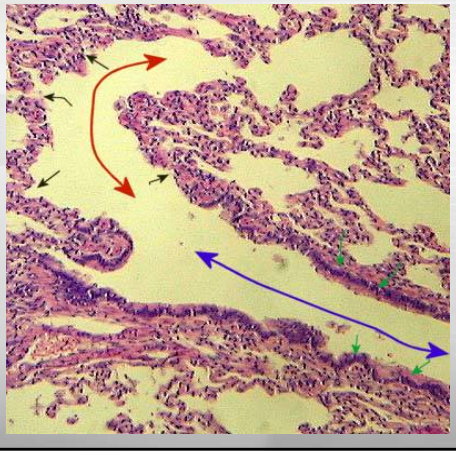
- X-linked lymphoproliferative disease

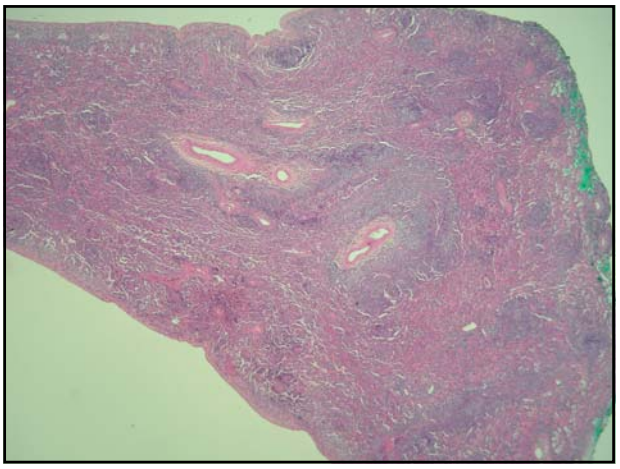
→gene testing of SAP/SH2D1A negative

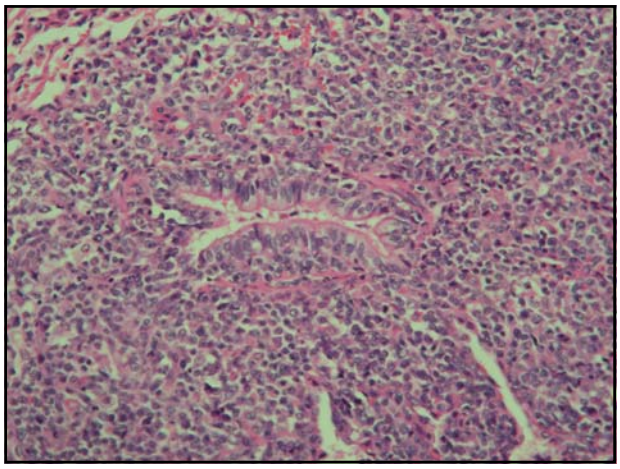
- Hemophagocytic lymphohistiocytosis

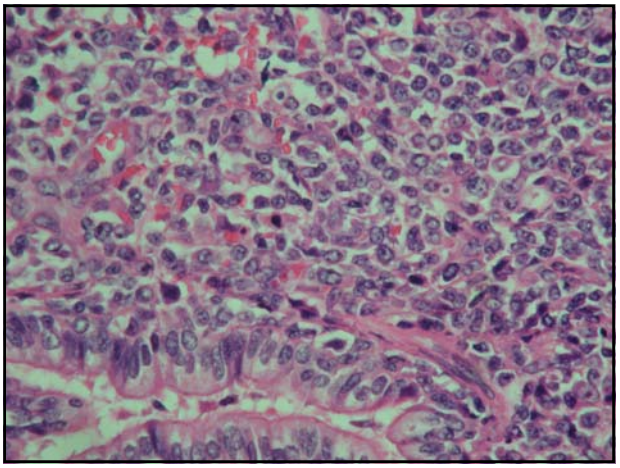
→bone marrow survey negative

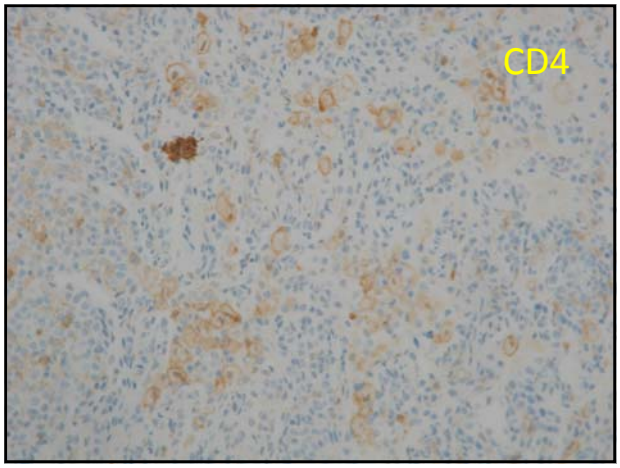
- Lymphomatoid granulomatosis

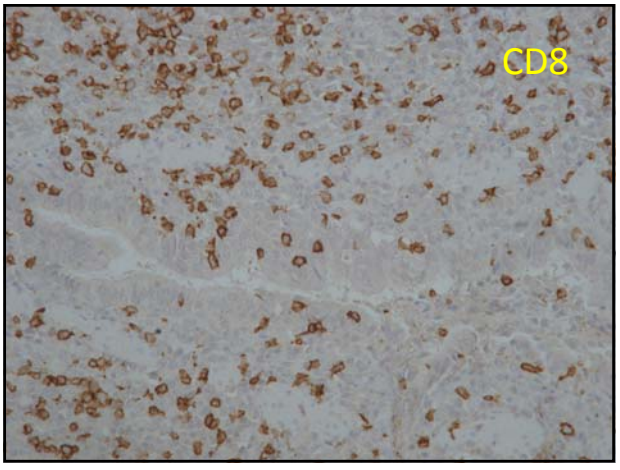


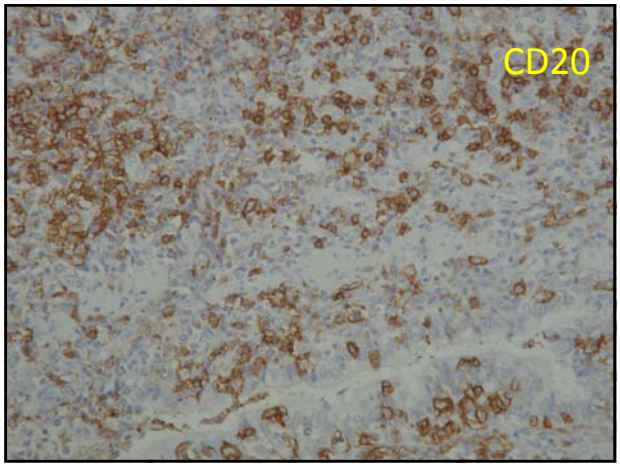


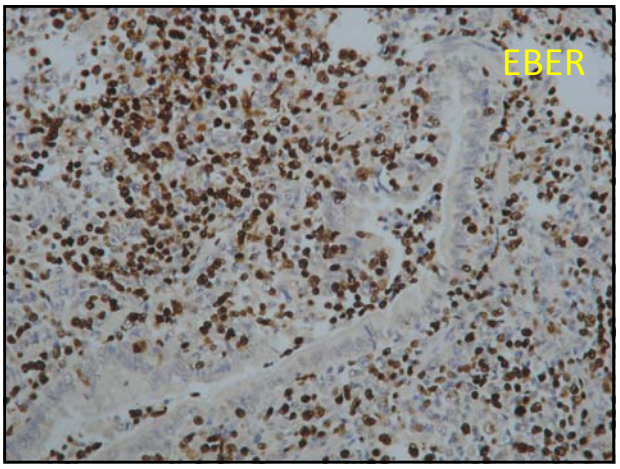





CD4


CD8


CD20




- Comaptible with lymphomatoid granulomatosis, grade III

## Disease progression

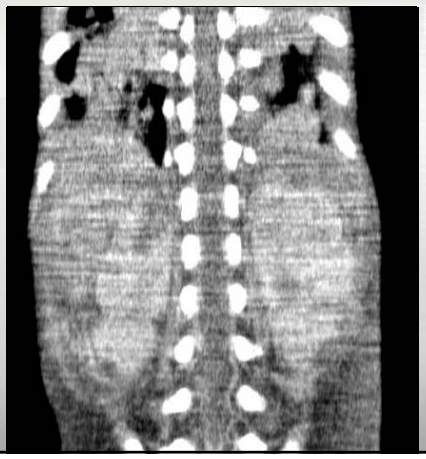
- Exposure keratitis(ou) and severe chemosis(os)
- Acute hepatitis

→r/o multiorgen involvement

- Rituximab once per week since 12/31
- Arrange whole body CT

## Whole body CT

- Multiple organ involvement of lymphomatoid granulomatosis including bilateral eyeballs, lung, liver, spleen, kidney, mediastinum, GI tract and brain.



- The patient expired on 1/15 due to cardiopulmonary failure despite of CPCR.

## Impression

1. LYMPHOMATOID GRANULOMATOSIS, GRADE III. status post Rituximab treatment, with multi-organ involvement
2. Pneumonia, favor CAEBV pulmonary involvement
3. Gastroesophageal reflux
4. Cardiopulmonary failure
5. Expired at 10:25 on 1/15

- Discussion

- Lymphomatoid granulomatosis is a rare EBV–associated systemic angiodestructive lymphoproliferative disease.
- It is characterized by prominent pulmonary involvement but can also involve multiple extrapulmonary sites.

- Lymphomatoid granulomatosis usually is progressive and fatal. Mortality rates range from 63-90% at 5 years
- The male-to-female ratio of lymphomatoid granulomatosis is 2:1.
- most common after the fifth to sixth decade of life

- Pulmonary involvement
- Skin: Patchy, occasionally painful, erythematous macules, papules, and plaques
- 25% patients had extensive lymphocytic infiltration of the meninges, cerebral vessels, and peripheral nerves
- Renal involvement
- Liver involvement

## Causes

- Other than its association with opportunistic disease and EBV, the etiology of lymphomatoid granulomatosis is unknown
- The cause of death is usually extensive destruction of the pulmonary parenchyma, resulting in respiratory failure
- Poor prognostic indicators :

  <30y/o ; neuro or liver involvement ; leukopenia

## Treatment

- Gancyclovir
- Rituximab:

  A monoclonal antibody that targets the B-cell surface molecule CD20

- Thank you for your attention