



PAEDIATRIC CASE 1

UMMC

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MRA

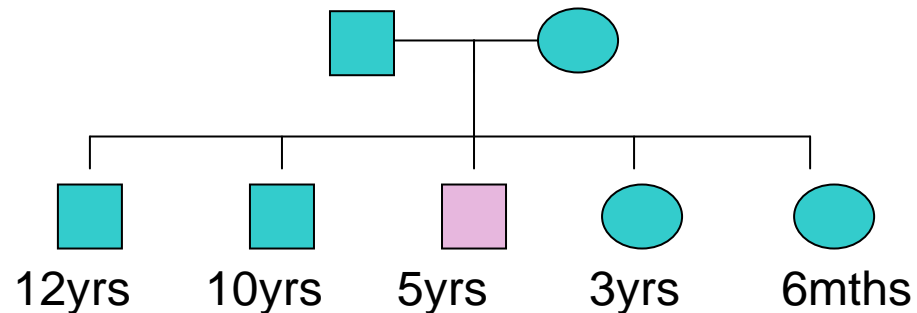
Worsening pleural effusion requiring ventilatory support



MRA

- 5 year old Malay boy
 - Non-purulent cough
 - Worsening dyspnoea
 - Loss of appetite and weight x 1 month
- } 2weeks
- No fever
 - No other associated symptoms
 - No history of contact with TB.
 - No history of trauma

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- Born term. Spontaneous vaginal delivery.
 - Birth weight: 3.3kg
 - Non consanguineous parents.
 - No significant family history
 - Immunisation complete. BCG scar present





Referred to Klang Hospital:

- Worsening respiratory distress
- Massive right pleural effusion
- Intubated and chest tube inserted



Investigation

FBC: Hb 12.7 g/dL
TWC $13.9 \times 10^9/L$ (Neut 44%)
Platelet $34 \times 10^9/L$

ESR: 9mm/hr

Albumin: 18g/L

INR: 2.16 PTT: 38.8s

Urine protein: Negative

Liver enzymes and renal function: Normal



Pleural fluid:

Appearances:	Haemoserous
LDH	533 IU/L
Protein	1417 g/dL
FEME	numerous pus cells
Cytology] negative
Culture	
AFB	



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- Mycoplasma serology Negative
 - Tracheal aspiration
 - Culture/ AFB/ PCR AFB Negative
 - Retroviral screen Negative
 - Blood/ Urine culture Negative
 - Mantoux Negative



CT Thorax and Abdomen (Klang)

- Massive (R) pleural effusion with hydropneumothorax
- Collapse consolidation of (R) lung with aerated upper lobe.
- Extensive consolidation (L) lower lobe with aerated upper lobe showing increased areas of attenuation with thickened septa.
- Significant pericardial effusion
- Minimal ascites
- Liver/Spleen/Pancreas/Kidneys normal
- No lymphadenopathy



Problems

- Pneumonia with massive right pleural effusion
- Hypotension secondary to volume losses
- Renal impairment
- Hypoalbuminaemia
- Coagulopathy
- Liver impairment



In UMMC at day 21 of illness

- Blood pressure stabilized with fluid replacement
- Less oedematous with albumin infusion.
- Renal and liver impairment improved gradually
- Coagulopathy improved with Vit K and coagulation factors replacement mainly FFP

However..


- Continuous pleural fluid loss up to 1.5 to 2L per day





Investigations

- FBC : Hb 13.5g/dL
TWC 20.1x 10⁹/L (N87%, L5%)
Platelet 215x 10⁹/L
- RFT : Urea 12.2mmol/L
Creat 139umol/L
- LFT : Albumin 15g/L
Bilirubin 26umol/L
ALT/AST 524/808 IU/L
ALP/GGT 61/64 IU/L
- PT/INR: 2.33/2.5

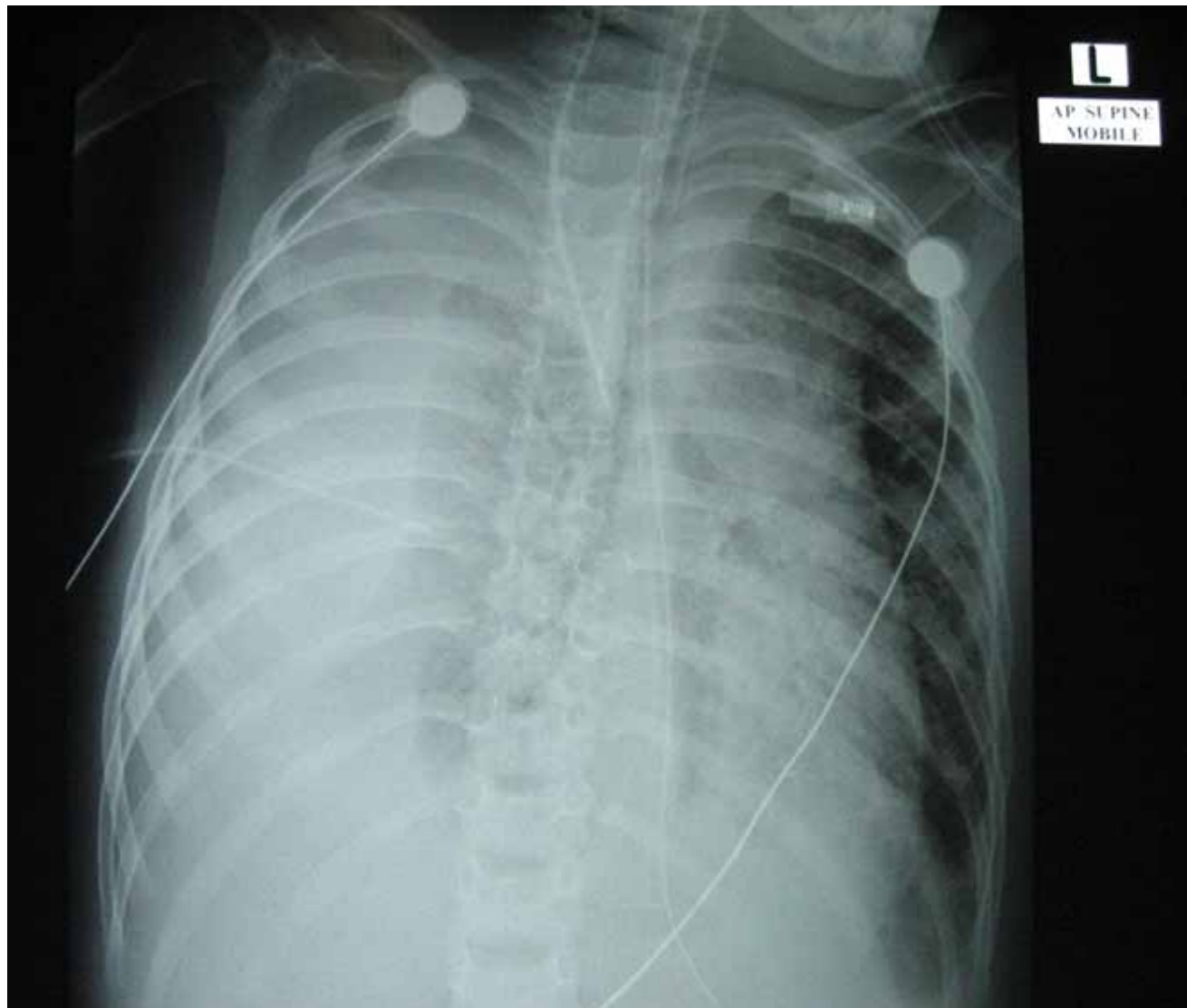
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- CRP <0.4
 - Cultures Negative
(Blood/Tracheal secretion/Pleural fluid/Peritoneal fluid)
 - Serum LDH: 247 IU/L

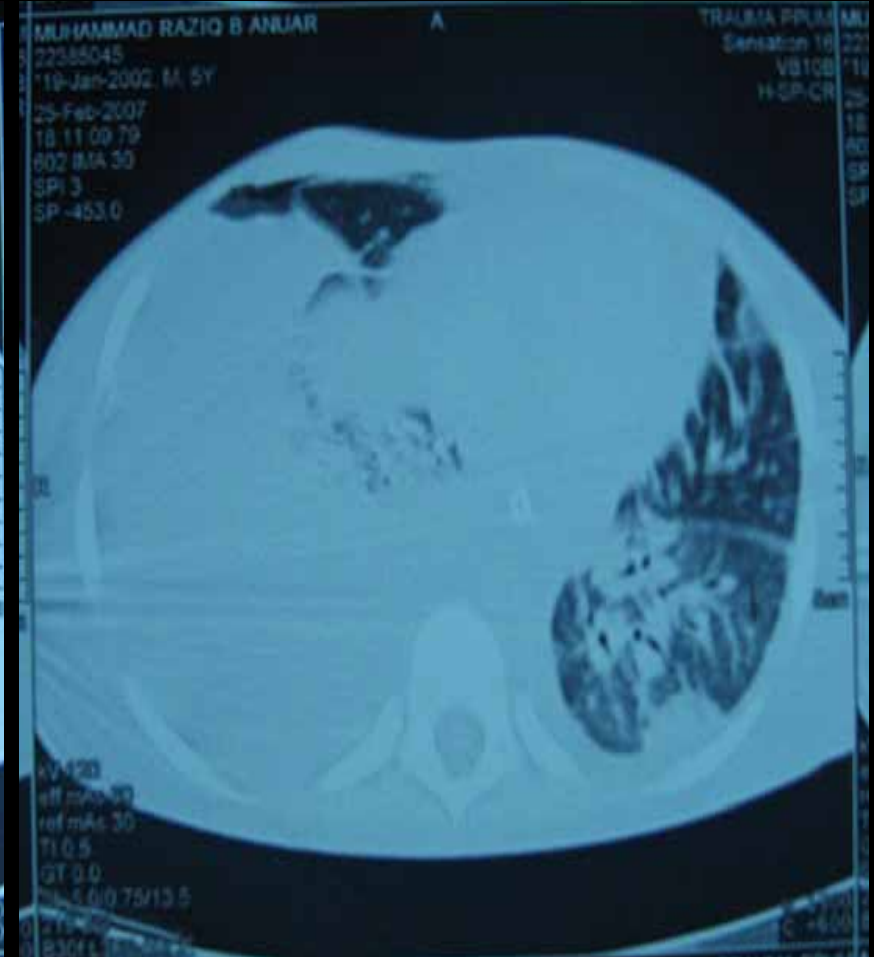


Pleural fluid

Appearance:	Haemoserous
RBC:	8800/uL
WBC:	370/uL
Polymorph:	7%
Lymphocyte:	93%
Protein:	41g/L
(Pleural/Serum Protein = 1.1)	
Organism:	Negative
LDH :	7 IU/L
Triglyceride:	1.6mmol/L

Admission



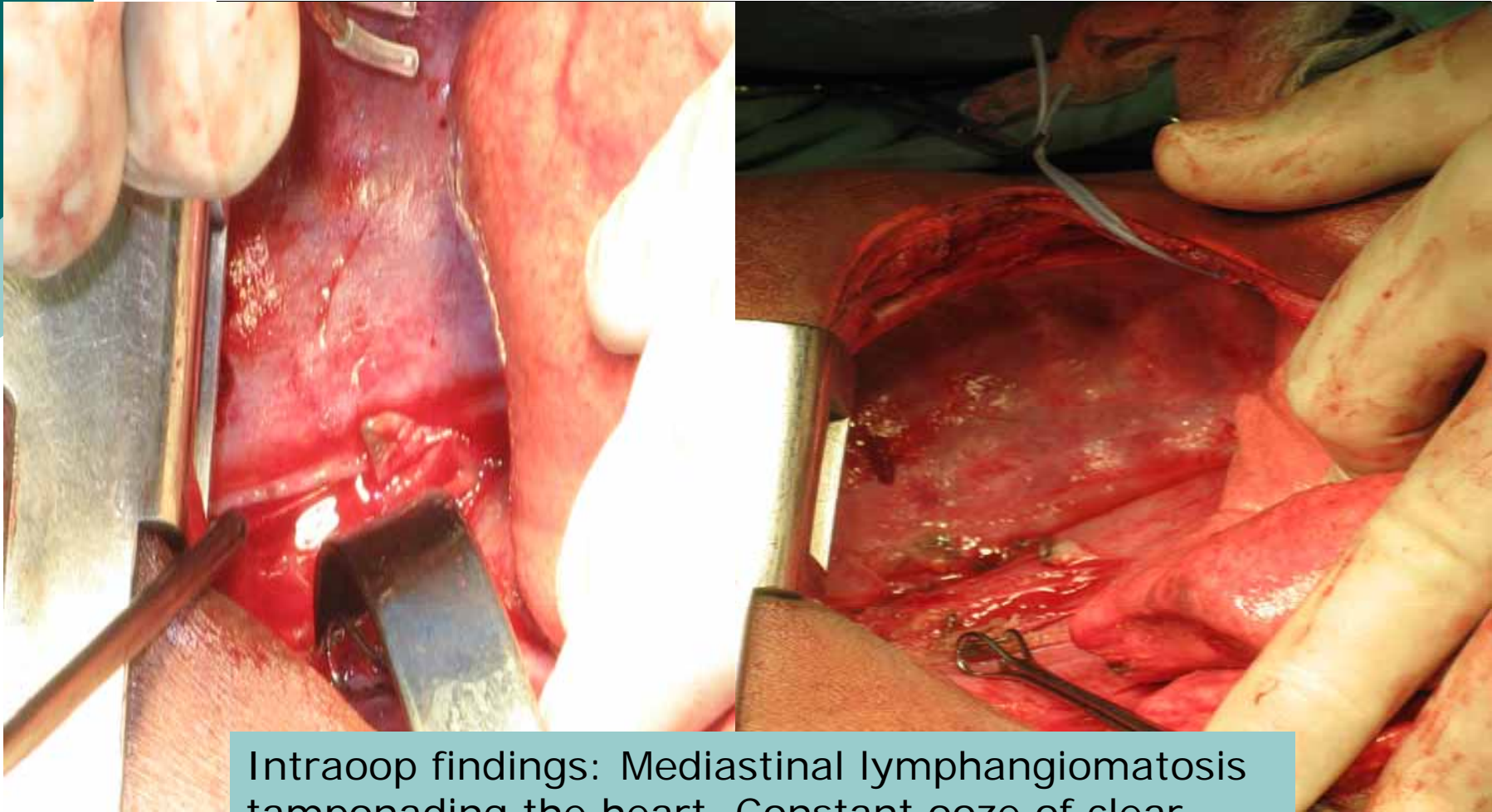




Working diagnosis..

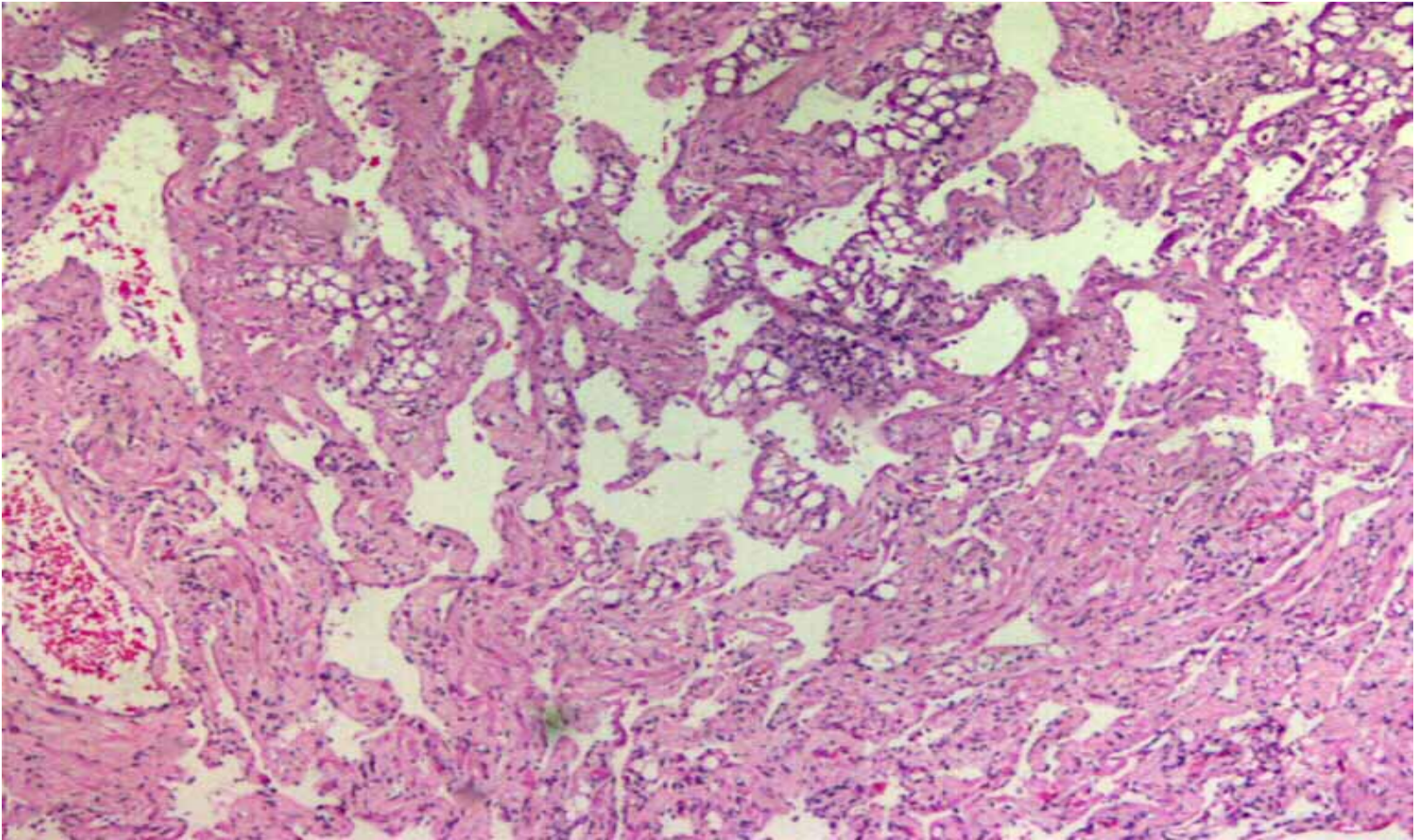
- ?Lymphangiomatosis
?Lymphatic injury
- ?Vascular injury
- ?Haemangioma
?Haemangiolympangioma
- ?Lymphoma

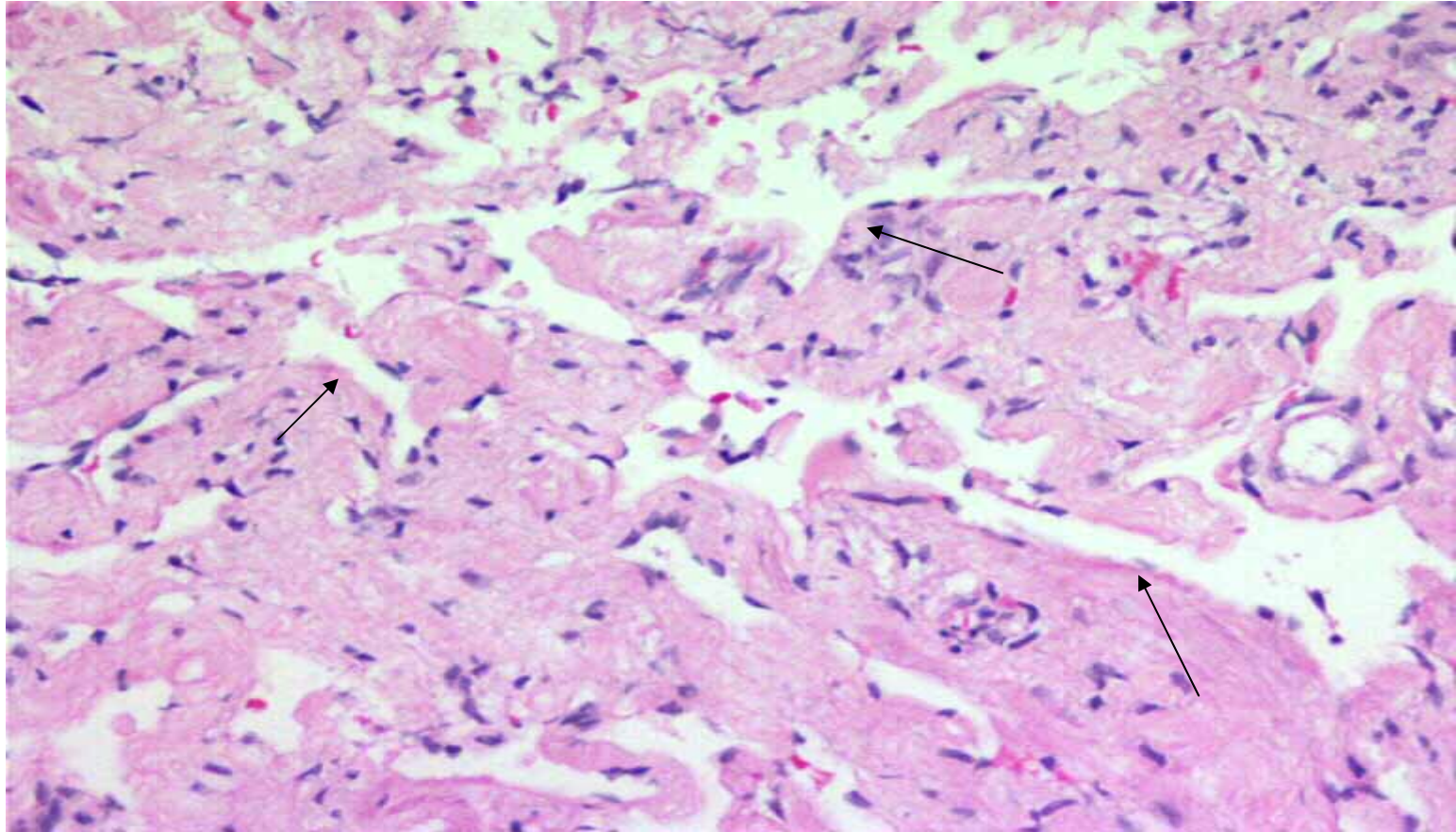
Surgery- (R) Thoracotomy, insertion of thoracoabdominal shunt.



Intraop findings: Mediastinal lymphangiomatosis tamponading the heart. Constant ooze of clear fluids. Thoracic duct intact

HPE



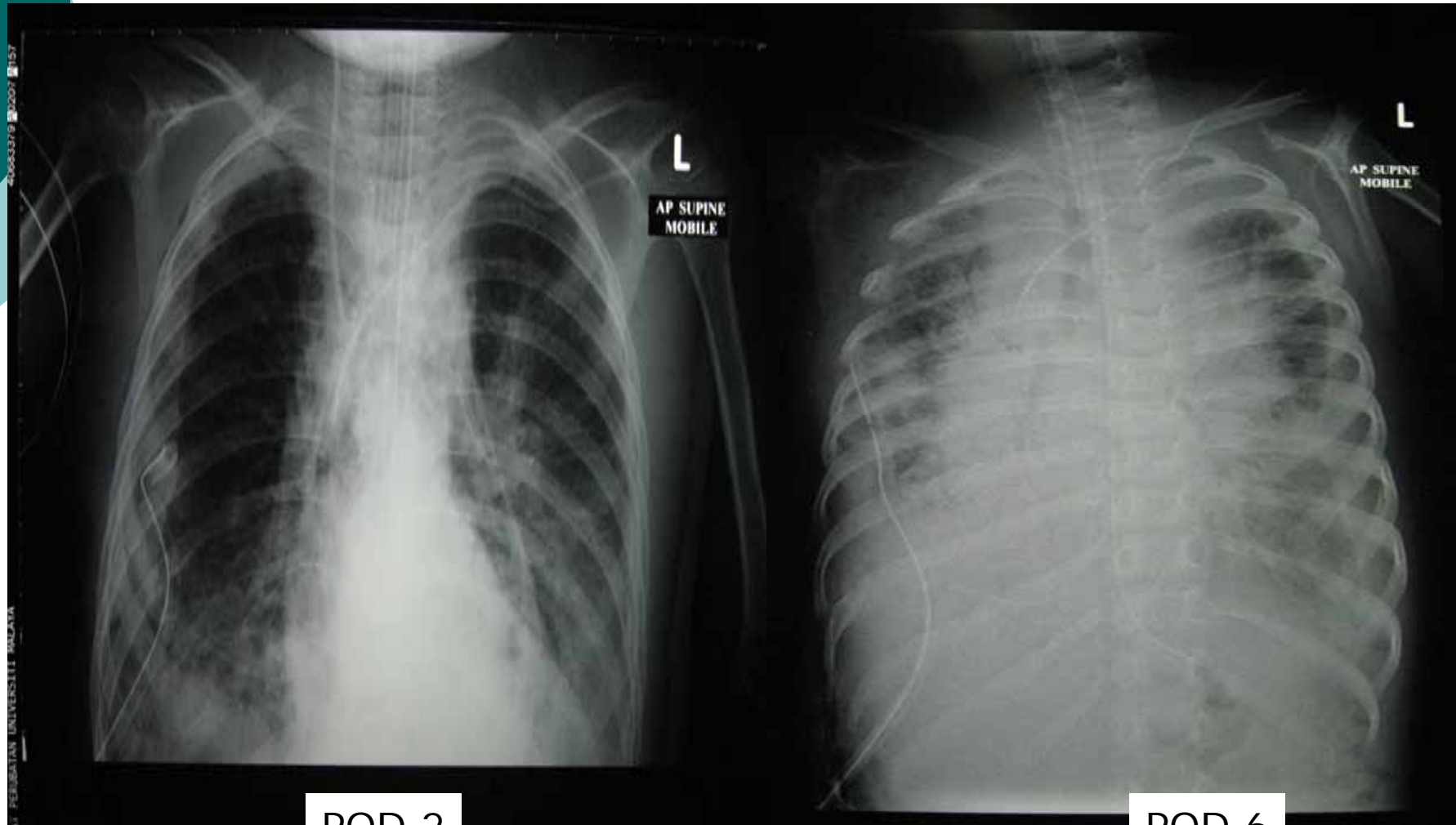


Fibrovascular tissue with many thin-walled channels compatible with lymphangiomatosis



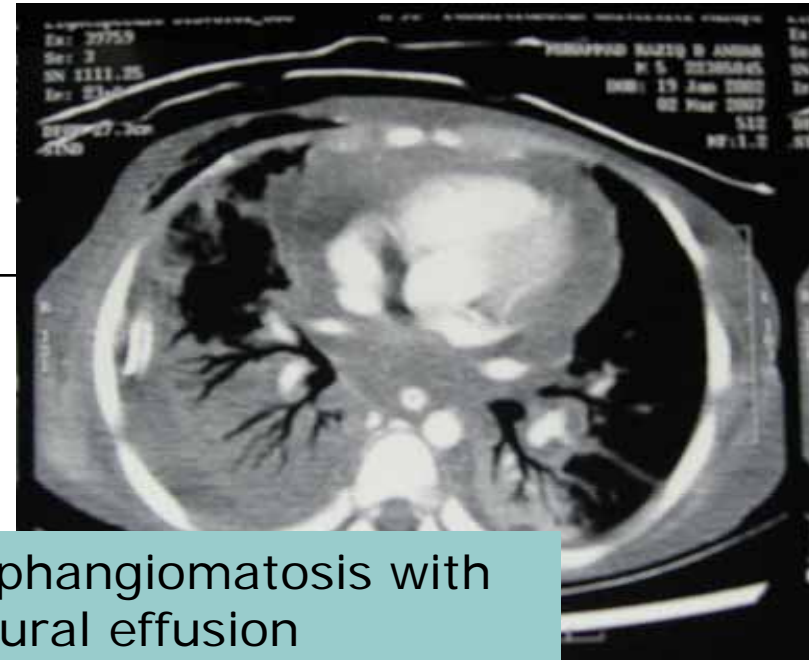
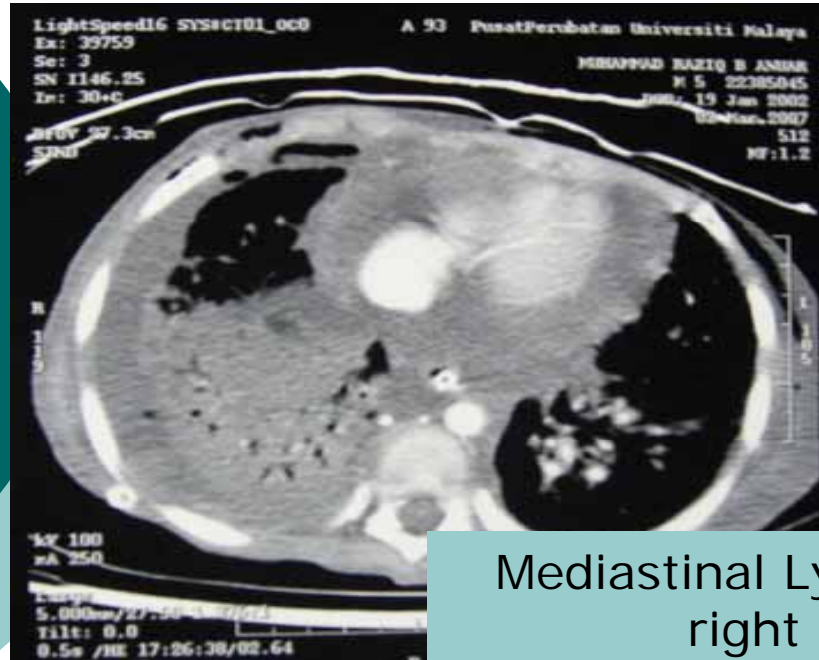
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- Started on SC Interferon at POD 3
1.5MIU daily ($1\text{MIU}/\text{m}^2$) then
increased at day 5 to $3\text{MIU}/\text{m}^2$

Post operatively

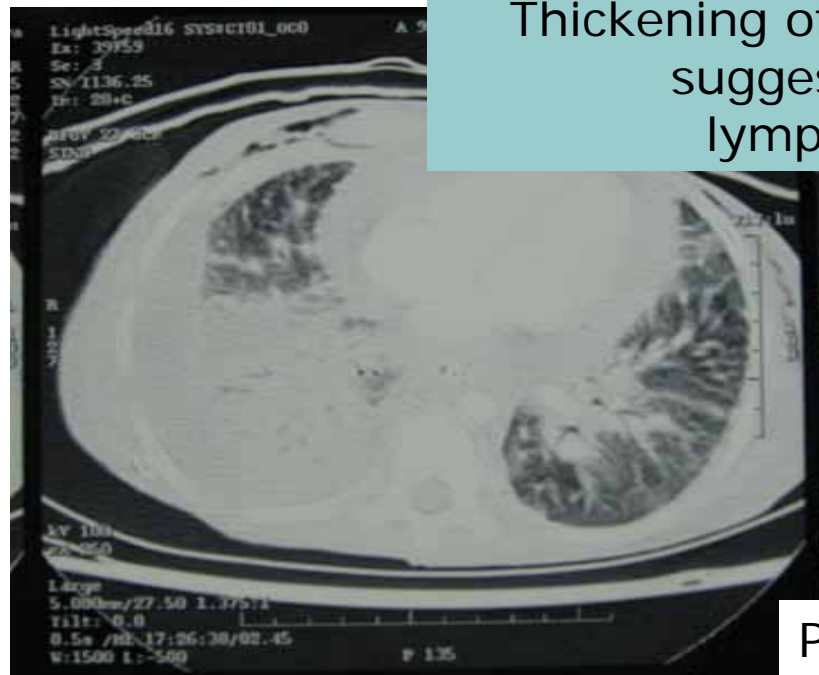


POD 2


POD 6



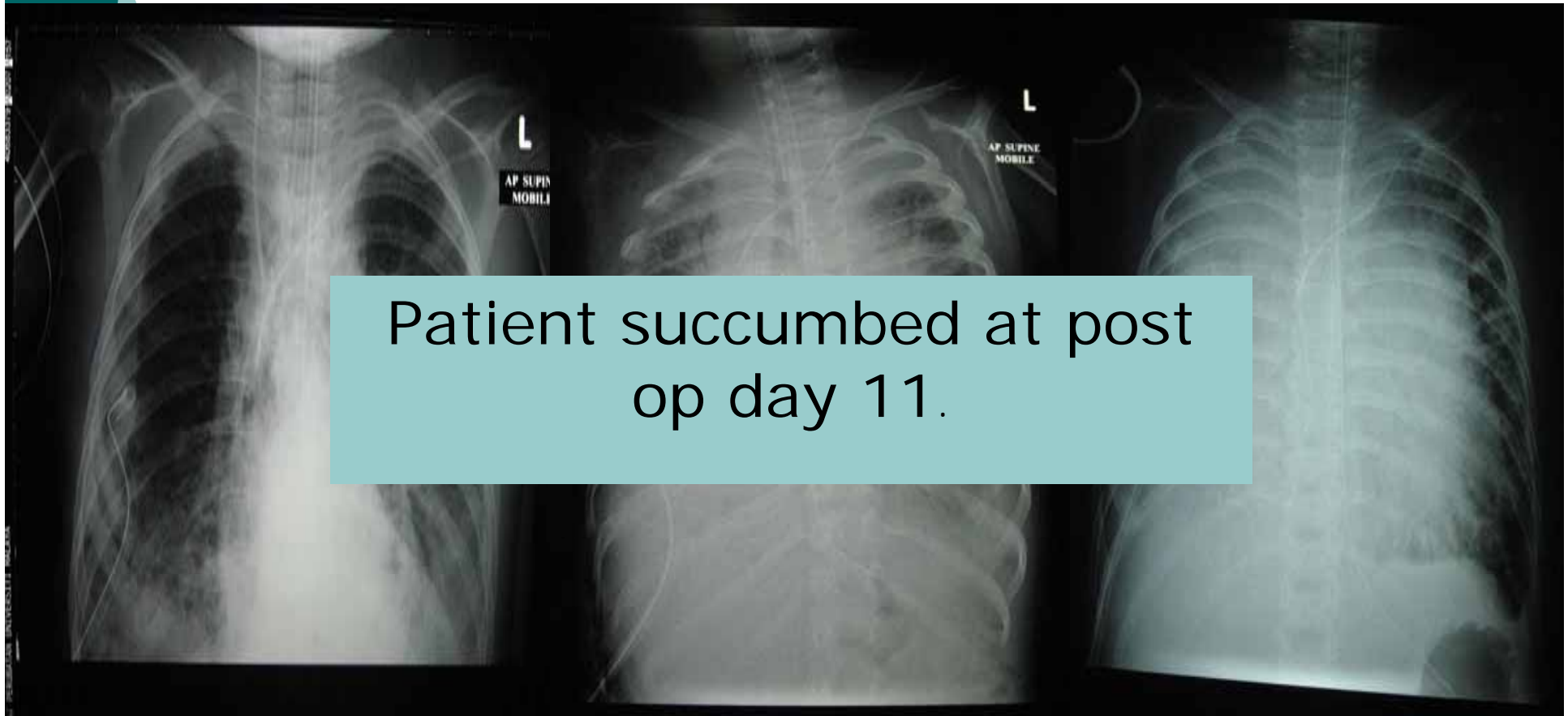
Mediastinal Lymphangiomatosis with right pleural effusion
Thickening of the interlobular septa suggesting pulmonary lymphangiomatosis



POD 6

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- Given first dose IV Vincristine 1.5mg/m² at POD10
 - Plan for IV Methylprednisolone 10mg/kg daily
 - Radiotherapy considered but patient too ill for transfer.

Post operatively



POD 2

POD 6

POD 11



LYMPHANGIOMATOSIS

- Rare congenital abnormality of the lymphatic vessels
- Mostly solitary and some associated congenital anomalies e.g. Noonan syndrome
- 0.7-4.5% of mediastinal tumours in children



LYMPHANGIOMATOSIS

- Involvement of multiorgan systems:
 - Musculoskeletal system (Gorham syndrome)
 - Lungs/pleura
 - Pericardium
 - Abdominal viscera
 - Testes
- Poor prognosis with chest and intra-abdominal involvement

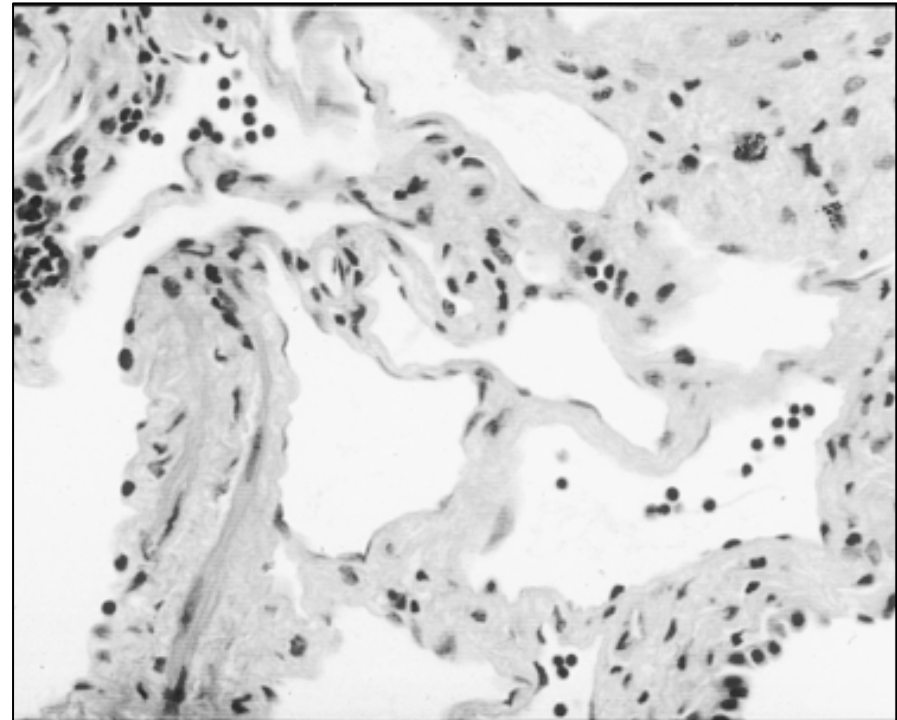


Presentation

- Asymptomatic – incidental finding
- Compressive nature
- Sequestration thrombocytopenia and consumptive coagulopathy

Pathologic features:

- Proliferation of lymphatics arranged as complex anastomosing spaces along anatomic lymphatic routes





Treatment

- Parenteral nutrition
- Medium chained triglyceride,
High protein diet
- Drainage of large fluid collections
- Sclerotherapy
- Surgical resection
- Systemic chemotherapy
- Interferon a
- Radiotherapy (18-20 Gy)

TABLE 1. Cases of Thoracic Lymphangiomatosis

Ref.	Age, Sex	Involvement	Treatment	Outcome, Follow-Up
2	Premie	Neck, scalp, lung, Pe	Respirator	DOD, 5 weeks
3	9 y, M	Med, lung, Pe, bone	Pleurocentesis	DOD, 6 m
	10 m, M	Neck, ax, Pe, spleen	Pleurodesis, pleuroectomy	DOD, 8 m
	11 y, M	Pe, bone	Pleurodesis, pleuroectomy	Alive, 5 m
	9 m, M	Pe, skin, bone, spleen	Pleurodesis, pleuroectomy	DOD, 33 m
4	1 m, M	Lung, Pe, pericardium	Heart-lung transplant	Died, age 7
	11 m, M	Lung	Unknown	Alive, 2.5 y
	3 y, F	Lung	Unknown	Sxs, 8 y
	3 y, M	Lung, Pe	α IFN	DOD, 1.5 y
	5 y, M	Lung	Bronchodilator	Alive, 6 m
	7 y, M	Lung, Pe	α IFN	Alive, 7 y
	16 y, M	Lung, Pe	Low-fat diet	Alive, 1.5 y
	20 y, F	Lung, Pe	Medroxyprogesterone	Alive \downarrow PFT, 13 y
	33 y, M	Lung	Tamoxifen $\times 2$ y	Improvement
5	20 y, M	Med, lung, RP, DIC	Observation	Alive, 1.5 y
9	11 y, F	Lung, pericardium, bone	Resection	DOD
10	1 y, M	Lung, pericardium, spleen, DIC	α IFN, P, low-fat diet, VCR tamoxifen, splenic embolization	Alive
11	20 m, M	Neck, ax, spleen, bones, DIC	Resection, VCR, splenectomy	Alive
12	4 m, M	Mesentery, RP, spleen bone, DIC, ascites, Pe	Partial splenic embolization	Recurrence Pe DOD 10 m
18	21 y, M	Sc, med, ax, bone	Resection $\times 2$, XRT	Alive
19	20 y, F	Lung, Pe, pericardium	Medroxyprogesterone $\times 4$ y	Alive
20	36 y, M	Hilum, RP, mesentery, bone	Resection, pleuro and pericardiocentesis, XRT	Alive
21	10 y, F	Med, sc, skin	XRT	Well, 8 y
	21 y, M	Pe, med	XRT	Well, 32 m
	13 y, M	Med, bone, liver, spleen, bladder	XRT	Well, 20 m
22	59 y, F	Neck, med, bone, liver, spleen	Resection	Improved, 3 m
23	3 y, M	Lung, spleen, bones	α IFN $\times 28$ m	Alive

24	NB, M	Lung	Resection	Unknown
25	27 y, F	Med, paratracheal	Resection	Doing well
	44 y, M	Med	Resection	Doing well
	51 y, M	Med, pericardium	Unknown	Symptomatic
	63 y, M	Med, paratracheal	Resection	Doing well
	3 y, F	Med, neck	Resection	Recurrence, 3 y
	8 y, F	Neck, paratracheal, lung	Unknown	Unknown
	14 y, M	Med, neck	Unknown	Asymptomatic
	27 y, M	Paratracheal, neck	Unknown	Recurrence
	4 y, F	Med, neck	Unknown	Unknown
	42 y, F	Paraspinal, Pe, bones	Resection	Recurrence Pe
	31 y, M	Med, flank	Resection	Recurrence Pe
	76 y, M	Paraspinal	Unknown	Asymptomatic
	68 y, F	Paraspinal	Unknown	Unknown
	30 y, M	Med, pleura, Pe, bones	Unknown	Unknown
	26	28 y, F	Med, Pe, bone	Pleurocentesis, ligation of thoracic duct, low-fat diet
27	NB, M	Pe, lung, skin	Respirator, pleurocentesis, low-fat diet	Alive, 1 y

28	9 y, M	Pe, lung, pleura, bone	Pleurocentesis, pleurodesis, low-fat MCT diet, steroid, diuretic, chlorambucil	DOD, 6 m
	10 m, M	Pe, pleura, lung, spleen, pericardium, bone	Pleuroectomy, pleurodesis, low-fat MCT diet	DOD, 4 m
	11 y, M	Pe, pleura, lung, bone	Pleuroectomy, pleurodesis, low-fat MCT diet	Resolved, 4 m
	9 m, M	Pe, pleura, skin, spleen, bone	Pleuroectomy, low-fat MCT diet, ligated thoracic duct, pleurodesis	DOD, 3.5 y
29	11 y, M	Pe, lung, pleura	Pleurocentesis, pleurodesis, XRT	Alive, 2 y
30	12 y, M	Pe, bone	Pleuroectomy, pleurodesis	Alive, 8 m
31	14 y, F	Noonan syndrome, Pe, chest, abdomen	Pleurocentesis, diuretic, steroid, α IFN	Alive, 1 y
	16 y, M	Pe, bone, spleen, paraspinal lesions	α IFN	Alive, >4 y
This report	8 y, M	Med, pleura, lung, Pe, bone, DIC	Pleurocentesis, diuretic, P, α IFN, EACA, XRT, VCR, tamoxifen	DOD, 3 m

α IFN, alpha interferon; ax, axillary; DIC, disseminated intravascular coagulation; DOD, dead of disease; EACA, epsilon-aminocaproic acid; MCT, medium-chain triglyceride; med, mediastinum; NB, newborn; P, prednisone or corticosteroids; Pe, pleural effusion; PFT, pulmonary function tests; RP, retroperitoneal; SC, supraclavicular; Sxs, symptoms; VCR, vincristine; XRT, radiation therapy.



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THANK YOU