

Grand Round Case presentation

By

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Learning Objectives

- To enhance the awareness of all staff members of our hospital about the Diagnostic approach & problem solving in pulmonary medicine.
- To ensure that reaching the end line diagnosis for difficult cases which require multidisciplinary approach is possible in KFH , if we are determined.
- To emphasize the importance of patient education , counseling and sharing in decision making for the success of any treatment plan.
- To ensure that working as a team has the greatest role in achieving success.

Case history

- 33 year old unmarried saudi lady.
- Presenting complaint :
 - Non productive cough started 1 year before presentation .
 - 6 months later , she developed Breathlessness on moderate exertion, progressive .
 - It progressed gradually till it became very difficult for her to move around the house, or to reach the toilet
 - No orthopnoea , No PND.
 - No chest wheeze.
 - No haemoptysis.

- No variability of symptoms .
- No relation to dust , perfumes , dander , detergents or food .
- No reflux symptoms.
- No post nasal drip.
- No joint pains or skin rash .
- No fever or sweats .
- Loss of appetite & weight loss 10 kg in 6/12.

- ROS :
 - No CV , Neurological , GI or GU symptoms.
 - Menstrual H. Currently regular menses .
- Past Medical / Surgical H. :
 - Diagnosed with Hyperthyroid Grave's dis. 9 Years ago and was on Antithyroid drugs and B- Blockers till 3 years ago when she had a Subtotal Thyroidectomy , according to patient preference . Post op FU revealed normal TSH .
 - Had troubling eczema 15 years ago which required topical and systemic steroids which was also discontinued 8 Years ago as she improved .

- Medications :
 - No medications at the time of presentation.
- Allergies :
 - None at the time.
- Family H.
 - No similar condition in the family.
 - Half sister has RA .
 - 2 sisters with diabetes .
 - No h. Of TB in the family.
- Social H. :
 - Unmarried . Works as a part time school teacher.
 - Lives with parents , brothers & sisters. She receives enough support from her family.
 - No pet animals.
 - No recent travel or contact with any P.T.B. patients
 - Her illness has severely affected her activities of daily living . She became disabled by the dyspnoea and consequently felt low self esteem.
 - She is obviously concerned that she might have something nasty .

In Summary

- 33 year old unmarried lady that presented with a **1 year history of progressive non productive cough** followed , in 6 months , by :
- disabling dyspnoea on mild exertion .**
- She was treated for **Grave's dis** with good recovery after surgery.
- She was also treated for **Eczema** 15 years ago for 6 years which subsided afterwards.
- Family history of **B.A.** (half sister) and **vittligo** (2 sisters) .

What is your differential at this stage?

- DPLD** . (Interstitial Lung Dis .) including :
 - Idiopathic** or secondary to **C.T. dis .** , Granulomas (**sarcoidosis**)
- Airway hyper responsiveness.**
 - What is going with ?
 - What is against ?
- Vasculitis (Churg Strauss) .**
- P.Tuberculosis .**
- Malignancy**
- Any possibility for **Cardiovascular causes ??**
- PAH** (Primary pulmonary hypertension) .
- VTE**
- Any other suggestions ???

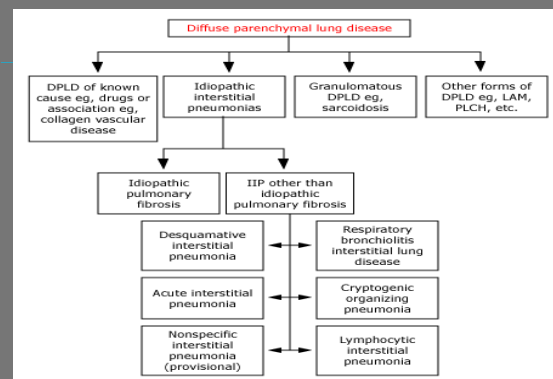
Examination

- She looked generally well at rest.
- Afebrile with normal other vital signs.
- Clinically Euthyroid.
- Thyroidectomy scar. No palpable Lymphadenopathy
- Chest Examination :
 - No clubbing , no Central cyanosis .
 - JVP not raised .
 - Trachea central .
 - Expansion symmetrically diminished.
 - Resonant percussion note bilateral.
 - Bilateral fine end inspiratory crackles** over both sub scapular regions , which did not change with cough.
 - The cough was a dry one
 - No rhonchi
- 6 minute walk test** could not obviously be performed as she was too breathless .

- CVS :**
 - Normal first and second heart sounds.
 - No added sounds .
 - No murmurs.
- Abdominal ex .** No organomegaly.
- Skin examination** No lesions found.
- Rest of body system examination** unremarkable .

Has the D.D. changed now ?

- Interstitial Lung Dis . (**Idiopathic**)
- or (secondary) to **sarcoidosis** , **C.T. dis.**
- OR
- Malignancy (with **Lymphangitis carcinomatosis**) .
- P.Tuberculosis**



What investigations shall we request at this stage ?

- Basic Bloods →
- CXR →
- HRCT chest. →
- PFTs →
- ABGs. →
- Immune screening →
- HIV test **NEGATIVE**
- TSH →
- F.O. Bronchoscopy & TBLB →
- Thoracoscopic Lung biopsy →

Basic Bloods

- WCC 7.9
- BUN 3.4
- AST 21
- ESR **75**
- S calcium 2.08
- Hb 13.9
- Crea 77
- ALT 12
- Bilir. 7/2
- CRP neg.
- Albumin 34
- Plt. 237
- Na 134
- k 3.6

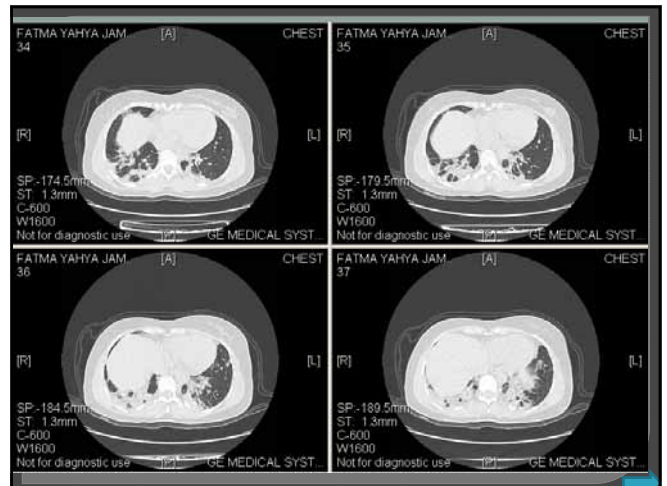
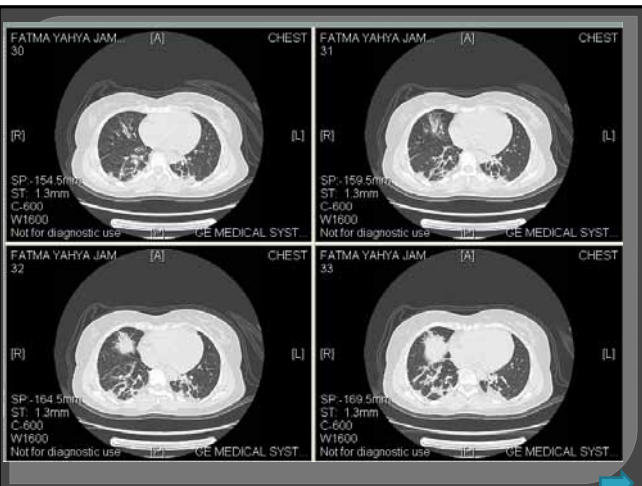
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ABGs

- Breathing air
- PH 7.43
- PO2 68
- HCO3 24
- PCO2 36
- O2 SAT 94 %



Immune screening

- ANA 1/80 then 1/40 on FU
- c ANCA < 3.1 (Ref. Range 0.0 - 10.0)
- p ANCA < 3.1 (Ref. Range 0.0 - 6.0)
- Anti SSA Ro Ab < 3.1 (Ref. range 0.0 - 4.0)
- Anti SSB La Ab < 3.1 (Ref. Range 0.0 - 4.0)
- Ig G 19.3 (N 8 - 18)
- Ig M 1.3 (N 0.6 - 2.5)
- Ig A 4.27 (N 0.9 - 4.5)



Thyroid screening

- TSH 14 (Ref. Range 0.27 - 4.2) uIU / l.
- T4 13 (Ref. Range 12 - 22) pmol /l.
- T3 3.8 (Ref. Range 2.8 - 7.1) pmol / l.



PFT's

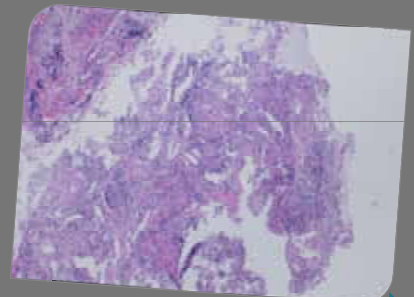
	Predicted	Best	% predicted
FVC	3.26	1.12	34 %
FEV1	2.74	1.00	36 %
FEV1/FVC	0.84	0.89	107 %
FEF 25-75	3.10	1.29	42 %
PEFR	6.39	1.87	29 %



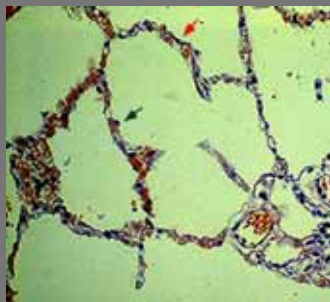
Bronchoscopy (BAL & TBLB)

- BAL cytology :**
- Endobronchial cells, numerous histiocytes and lymphocytes
 - Few neutrophils .
 - No malignant cells .

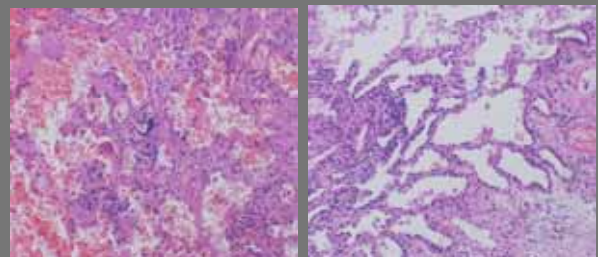
- TBLB :**
- Chronic inflammation and FIBROSIS.
 - Stains for Fungi & Mycobacteria are negative.
 - NO Granuloma .
 - No malignancy .



Normal Lung Histology



VATS



Thoracoscopic Lung biopsy (VATS)

- Lymphoid aggregates and fibrosis consistent with

LIP

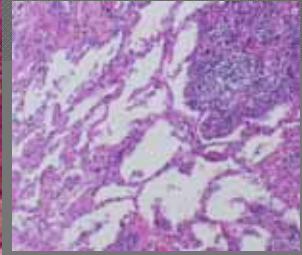
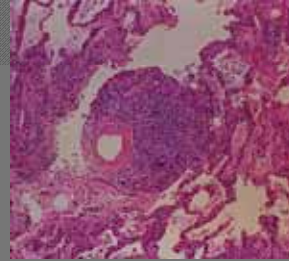
(Lymphocytic Interstitial Pneumonia)



Thoracoscopic Lung biopsy (VATS)

LIP MODEL FROM
UP TODATE

OUR PATIENT .



What is LIP

- LIP is one entity within a spectrum of lymphoproliferative disorders that can involve the lung.
- LIP represents a benign polyclonal proliferation (usually of mature B cells)
- An autoimmune etiology for LIP is attractive given its association with autoimmune processes
- LIP may predate the establishment of the protein abnormality or other diagnoses

- Sjögren's syndrome is associated with one fourth of reported cases of LIP.
- The pulmonary disease may proceed or follow the diagnosis of the underlying process.
- marked increased incidence in persons infected with the HIV virus, especially children [11-13]
- significant increase in the presence of EBV genome compared to other chronic lung disease specimens [19]

- It is particularly important to establish the benign nature of the lesion.

• C.P.

- Cough (71 %)
- Dyspnoea (61 %)
- Weight loss (16 %)
- Fevers (10 %)
- Pleuritic chest pain (6 %)
- Fatigue
- Arthralgias

Treatment

- Patient was fully educated and counseled about treatment including duration and side effects.
- Oral steroids started according to protocol adopted by the ATS at 1 mg / kg /day for an initial 8 weeks .
- Bone prophylaxis was also started .

Follow up to assess response

After 2 months of continuous monitored high dose oral prednisilone 45 mg per day

Clinical Response

- Cough disappeared completely .
- No more dyspnoea on effort (jumped from NYHA class III to normal exercise tolerance).
- **6 minute walk test**
- **Patient could walk briskly for 1140 meters (1.14 km) on flat ground .**
- A major positive switch of the patient's mood and self esteem
- **Minimal cushingoid features.**

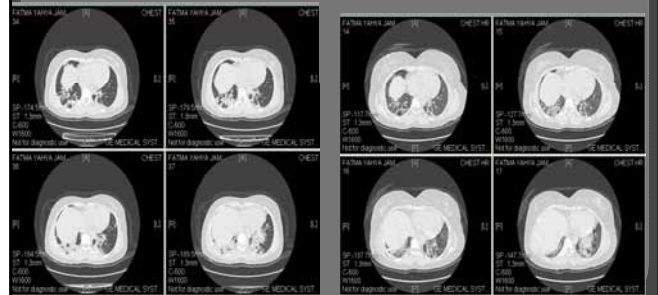
Laboratory changes

- **ESR**
 - Dropped from **75** prior to treatment
 - To **30** 2 months in treatment.
- **PFTs** planned shortly in Jeddah as they are not available here.

Radiological Response

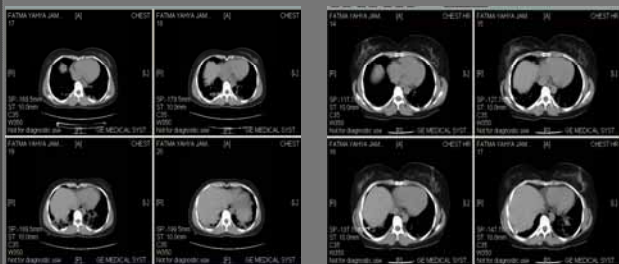
BEFORE TREATMENT

2 MONTHS IN TREATMENT



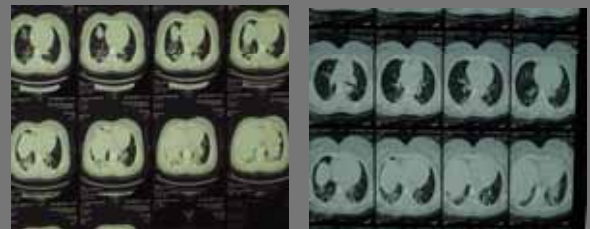
BEFORE TREATMENT

2 MONTHS IN TREATMENT



BEFORE TREATMENT

2 MONTHS IN TREATMENT



Plan and prognosis

- To gradually taper steroids to maintenance dose of 0.25 mg / kg and continue this dose for 1 year.
- Consider the possibility of **relapse** after stopping steroids (strict F.U. is required).
- Consider that a small fraction of those patients **develop Lymphoma** (usually low grade & slowly growing)
- **Prognosis :**
 - 52 % with good response .
 - 11 % remain stable.
 - 37 % deteriorate (median survival of 20 months) .

Lessons and take home messages from this case

- Good communication with patients makes all the difference.
- Give the patient understanding and empathy.
- Spend time with your patient to explain fully what you are doing.
- Early treatment cannot be overemphasized for successful outcome.
- Let the patient take informed decisions , and share the decisions with you.
- If we have the knowledge , confidence and the ability to perform as a team , we can do as good as the best centre in the world.



Thanks a lot for interacting

Have a nice day.