Pyrexia under investigation with severe cough in an immunocompromised host

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History

- 33 year old female housewife presented with a history of cough for 1 month. Cough was associated with low grade evening fever, loss of appetite and loss of weight for 1 month.
- She is a known case of inflammatory bowel diseasefor 15 years and she is on sulfasalazine on regular basis, with intermittent use of steroids during flare. She is also hypertensive on telmisartan 40 mg daily since 5 years.

- Past history of hospitalization for acute bronchitis, acute gastritis and respiratory distress in July 2022.
- She was seen by gastroenterologist, pulmonologist, critical care physician
- Multiple blood investigations were done suggestive of raised CRP, WBCs, ESR
- X ray Chest showed ill defined opacities being treated as pneumonia with antibiotics, and bronchodilator without any relief

- What do you think what is your next step?

Examination

- On Examination: T- Normal.

  Pulse – 100/min

  Blood Pressure- 120/70 mmhg

  RR- 14/min

  RS- clear

  Sp02- 99%

  Per abdomen- soft

- Which investigations will you like to do?

Investigation

- On Blood investigations: Hb- 8.4, TC- 16,200, PC- 644000, Eosinophil- 0.7%, CRP-28.1, SGPT- 42, S. Creatinine-0.6, ESR-90, Urine routine and micro- Protein present, Pus cell 8-10, RBC- plenty.
- On repeat hemogram Hb-8.23,TC-10,100, PC-103300, Eosinophils- 0.2%, RBS- 92.98, S. Creatinine- 0.6, HIV and HBsAg negative.PT/INR- 20.1/16/1.29, APTT- 26.9
- USG abdomen- right lobe of liver hemangioma on July 2022.
- USG of neck – No lymphadenopathy on July 2022.
- CT chest showed Concentric thickening involving thoracic trachea on July 2022.

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USG ABDOMEN PELVIS:

Liver appears normal in size and echotexture. Portal vein is normal in caliber. No evidence of dilatation of intrahepatic biliary radicles or portal radicles.

Gall bladder is normally distended and appears unremarkable. Wall thickness appears normal. No calculus or SOL. CBD is normal in size. No evidence of stone in CBD.
Pancreas is normal in size and echogenicity. No evidence of any focal lesion. MPD is not dilated.
Spleen appears normal in size and echotexture.

Kidneys: Normal in size and shape. No evidence of calculus, hydronephrosis or mass lesion is seen. Cortical thickness and echoes appear normal.
Right kidney : 10.4 x 4.0 cms. Left kidney : 11.5 x 4.4 cms.

Urinary bladder is mildly distended and appears unremarkable. Wall thickness appears normal. No evidence of calculus, internal echoes or sol noted.
Uterus and ovaries appear normal.
No evidence of free fluid is seen in peritoneal cavity.
No evidence of para-aortic lymphadenopathy is noted.

POSITIVE FINDINGS :

- ***A small well defined round shaped echogenic soft tissue lesion of 8 mm in right lobe of liver – haemangioma.***
- ***Bilateral minimal renal cortical scarring.***
- ***No evidence of mesenteric-omental-bowel-IC junction thickening, lymphadenopathy, ascites or pleural effusion.***

DR. RAVI GAJJAR
Consultant Radiologist

Clinical Profile.- K/C/O spondyloarthropathy + IVD → C/o fever+ sore throat + coughing

MDCT (128 Slice) SCAN OF THORAX:

Technique:-
MDCT imaging was performed using sub-millimetre thin plain axial scan of thorax from thoracic inlet to diaphragm.

Findings:-
Evidence of mild concentric wall thickening is seen involving thoracic trachea, carina and proximal most main bronchi with perifocal mediastinal fat haziness. Maximum wall thickness along right lateral aspect including localized mediastinal fat strandings is about 4.7 mm. No evidence of discrete stricture formation is seen.
Multiple short axis centimeter-subcentimeter sized regional pre-para-peritracheal, perivascular and subcarinal nodes are seen.

Both lung fields appear clear.
No evidence of any significant interstitial or air-space abnormality is seen.
No pleural or pericardial effusion is seen.
Mediastinal vessels appear normal in calibre.
Visualised portion of unopacified oesophagus, thoracic cage and soft tissue appear unremarkable.

CONCLUSION:-

- **Mild concentric wall thickening involving thoracic trachea, carina and proximal most main bronchi with perifocal mediastinal fat haziness and regional-mediastinal nodes as described, -- possibility of underlying inflammatory nature can be more likely than infective. May needs onward investigation.**

What is your differential diagnosis?

- Bacterial infection
- Atypical infection due to immunocompromised status
- Tuberculosis
- Any other cause needing tissue diagnosis

Bronchoscopy for diagnosis

- Bronchoscopy showed diffuse thickening of mucosa covering trachea and both bronchial tree.
- No major secretions seen
- No cavitory lesions
- No Mass seen
- Tissue taken for HPE, genexpert, bacterial culture, fungal culture, AFB culture
- Genexpert, Bacterial culture were negative
- HPE report arrived…….

Histopathology report

Histopathology Report

Specimen :
Bronchial lung biopsy for HPE.

Clinical Diagnosis :
Diffuse thickening of trachea and bronchus. Known case of IBD, to rule out Amyloidosis
Ankylosis spondylosis.

Gross Examination :
Multiple fragments - 0.2 to 0.3 cm. Aggregate - 1x1 cm. All in 1 block.

Microscopic Examination :
Section shows bronchial, peribronchial & alveolar parenchyma with dense lymphoplasmacytic inflammation with lymphoid aggregates & few medium sized peribronchial illformed epithelioid granulomas with multinucleated giant cells.
No caseous necrosis seen.
No tissue eosinophilia seen.
N amorphus amyloid material seen.
No fungi/malignancy.

Impression :
Bronchial lung biopsy for HPE :

Moderate chronic bronchitis with prominent lymphoid follicles with illformed medium sized peribronchial epithelioid granulomas.

No caseous necrosis seen. No tissue eosinophilia seen.

N amorphus amyloid material seen. No fungi/malignancy.

Granulomas are not classical of mycobacterial etiology.

Above findigs granulomatous bronchitis may be seen secondary to Inflammatory bowel disease as a extraintestinal menifestation.

This is an Electronically Authenticated Report.

Diagnosis

- This a rare presentation of (Pulmonary) extra-intestinal manifestation of Inflammatory Bowel Disease !!!!!

Review

- IBD is associated with extra-intestinal manifestations(EIMs)that tend to parallel intestinal activity and have a debilitating effect on the quality of life.
- EIMs primarily affect the joints, skin, and eyes with less frequent involvement of the liver, kidney and pancreas.

- Extraintestinal manifestation of IBD.
  - Dermatological: Erythema nodosum, Pyoderma gangrenosum
  - Ophthalmological: Uveitis, Scleritis
  - Hepatic: Sclerosing cholangitis
  - Musculoskeletal: Spondyloarthritis

Pulmonary Manifestation of IBD

- Very few cases in literature
- Tend to be missed due to lack of tissue diagnosis being attempted
- Irrational antimicrobials are being emperically tried for long, after which patient gets exhausted
- This patient had intermittent lung complaints for 6 months on details history taking with worsening in one month
- Empirically mistreated as Pulmonary Tuberculosis in some scenarios

Treatment

- Educate the patient.
- Medical management will be steroids.
- Patient improved and achieved remission
- She was put on tofacitinib as steroid sparing agent, as she dint tolerate azathioprine and methotrexate
- She is currently stable

Take Home Message

- Every fever with cough is not necessarily an infection
- Immunosupressed host with fever and cough, usually can be any typical or atypical lung infection
- Whereever possible tissue diagnosis should be attempted
- Pulmonary manifestation of IBD, though a rare entity has to be kept in D/D of such patients who present with fever / cough and above radiological findings of CT Thorax
- High index of suspicion is needed to achieve appropriate diagnosis

THANK YOU