HLH and Acute Respiratory Failure in a young adult with disseminated TB :A Case Report

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A 34 year old lady, of Thai origin, who was recently diagnosed with Crohn's Disease, presented to the Emergency Department with bloody diarrhea ,dry cough and fever

She denies any recent travel history, but she was living at a women's Shelter at the time she came in to the hospital.

O/E:

1-General examination: GCS 15/15,cachectic,pyretic (39 C°), no lymphadenopathy

2-Respiartory and CVS examination: Unremarkable

3-Abdominal examination: Right lower abdominal tenderness with no rebound or rigidity.

Because we are currently in the pandemic ,multiple COVID swabs were negative peri hospital admission. Initial CT abdomen and pelvis was consistent with right sided colitis and terminal ileitis.

-Provisional diagnosis of flareup of Crohn's Disease was made , where she was medically optimized. She was started on high dose Steroids,Ocatasa and Adalimumab.

-However, she continues to have severe abdominal pain with bright red bleeding per rectum and worsening diarrhea. Second CT AP however showed stable appearances. Rescue Infliximab was then started with a negative QuantiFERON TB gold testing prior to commencement.

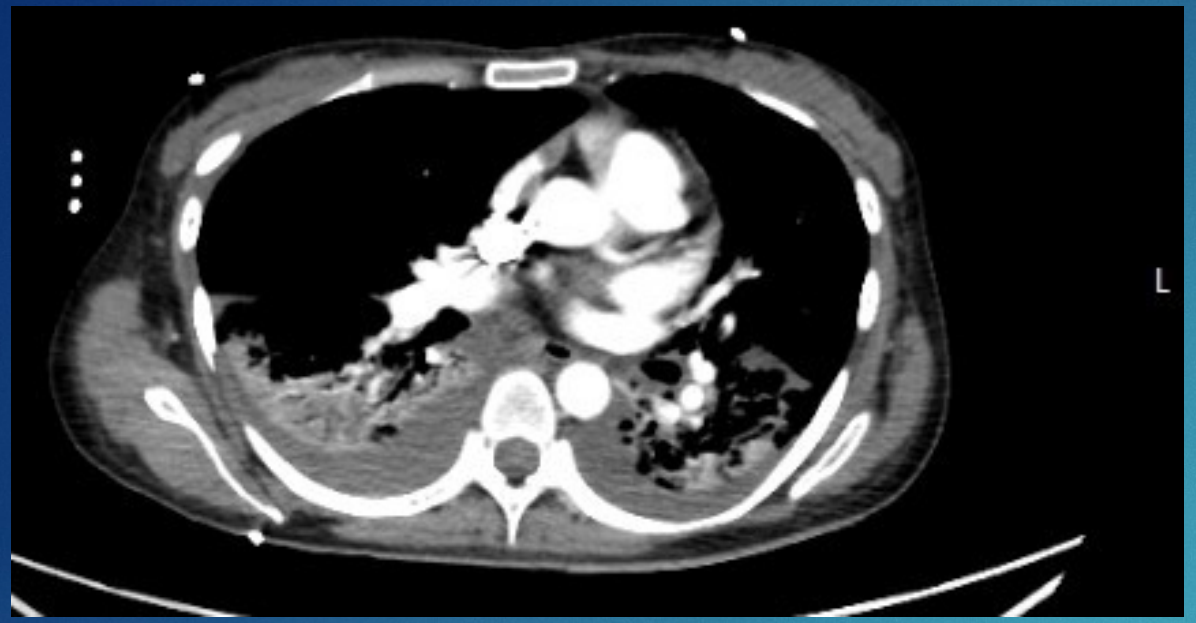
-Given the refractoriness to medical management even after immunotherapy, a decision to proceed with surgery was made after consenting the patient .

-She had Laparoscopic Right Colectomy with Diversion Loop Ileostomy (ileostomy formation in 1st stage and then reversal of ileostomy in the second stage) as anastomosis would be very risky.

-On the fifth day postoperatively, she became pyretic ,hypotensive and tachycardic. Bloods showed pancytopenia ,coagulopathy ,hyperferritinemia and severely deranged LFTs .

-Blood cultures at the time was negative for any growth.

-Further CT CAP was performed and showed Bilateral small pleural effusions with minor patchy ground glass opacities in the lung bases, reactive lymphadenopathy in the right abdomen and new multiple splenic microabscesses/microemboli.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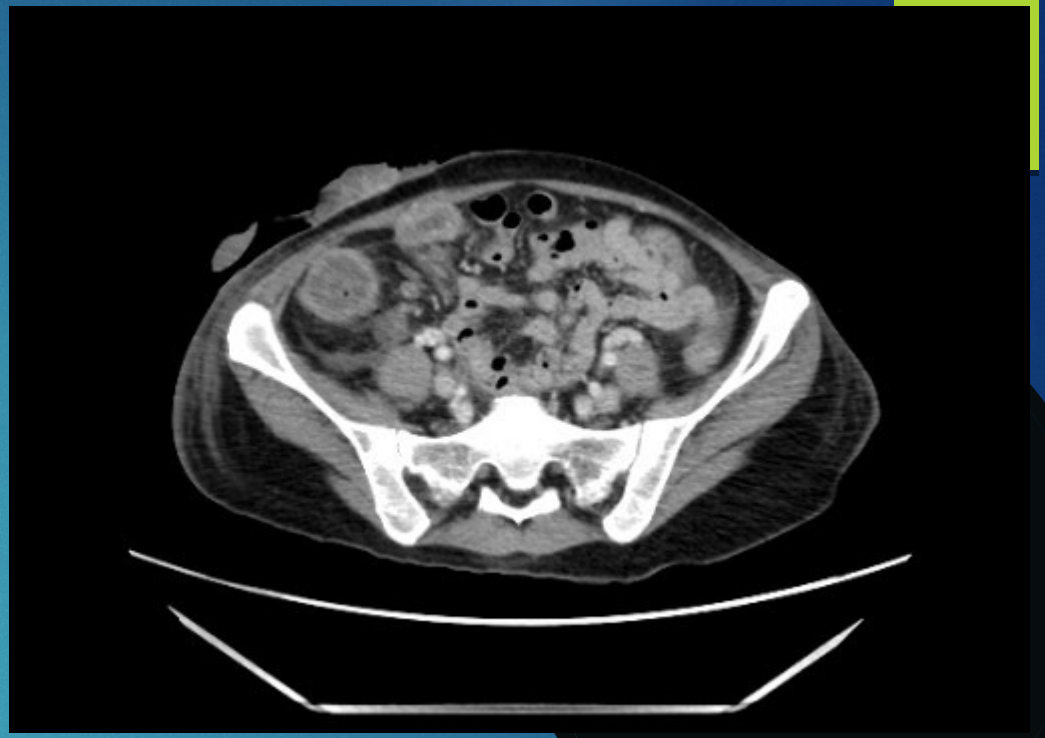
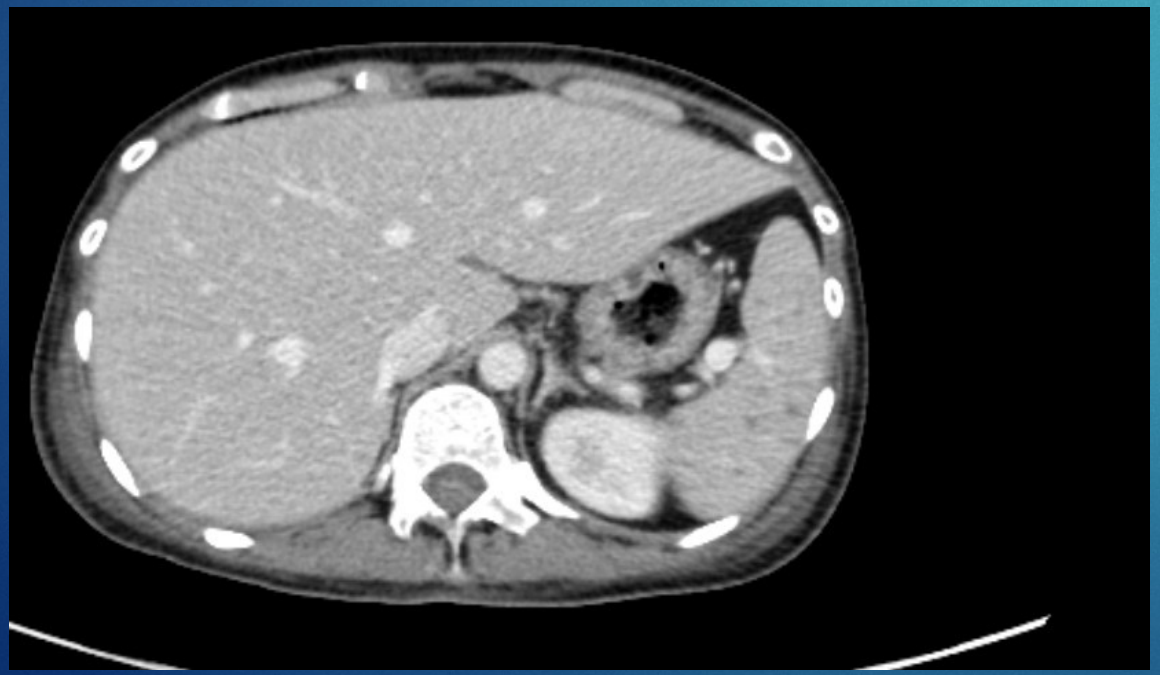




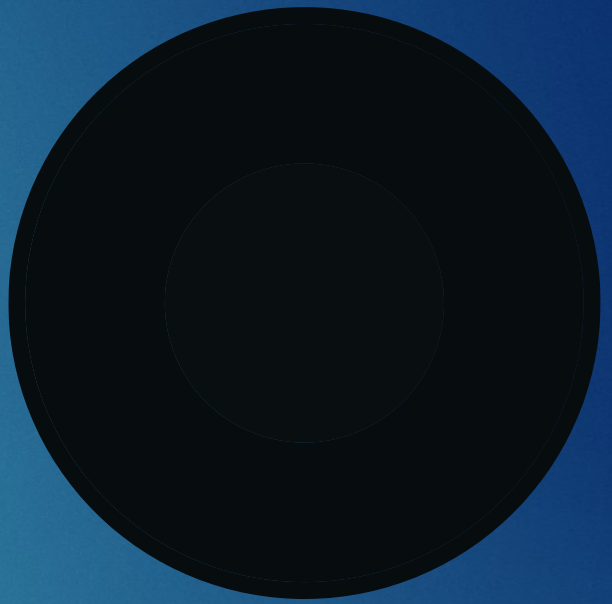
-A high index of suspicion for Hemophagocytic lymphohistiocytosis secondary to CD flareup /Refractory Sepsis.

-She was then transferred to the ITU due to increasing oxygen requirements for a short period of Optiflow, where she was placed on airborne precautions and treated as COVID Pneumonia/ARDS.

-Shortly after , she was intubated and ventilated . BM Biopsy confirmed Haemphagocytosis , whereby Anakinra ,IVIG and Dexamethasone are started with mild response. Etoposide was then commenced .

H score of the patient prior to the BM aspirate:

- Immunosupression - Yes +18
- Tem 38.4 - 39.4 +33
- Organomegaly - No
- Number of cytopenias - 3 +34
- Ferritin >6000 + 50
- Triglycerides 1.45 - 4 +44
- Fibrinogen <2.5 +30
- AST >30 +19
- BM Aspirate :awaited
- Score = 228 points (96 - 98%) probability of haemophagocytic syndrome.



-Although the clinical and radiological pictures were not compatible with Disseminated TB, the presence of new lymphadenopathy warrants the need to rule out TB, therefore bronchoscopic lavage / aspiration was sent for TB PCR, micro-culture including TB and fungal culture.

-2 days later, Bronchial washings were culture negative but trace positive for TB DNA with indeterminate Rifampicin resistance, which was unusual in that context for a disseminated disease not to be smear positive or at least fairly rapidly culture positive.

-She was started immediately on a liver sparing antituberculosis regimen ,(Amikacin, Moxifloxacin and Ethambutol) together with immunosuppressant withdrawal.

-The fast track PCR containing colonic biopsies taken prior to the surgery also came back also positive , with a further confirmation of a positive DNA probe of the BM aspirate .

-She was then successfully extubated together with improvement in her biochemical profile .She was transferred to a Respiratory Ward.

THANK YOU