A challenging case of hero syndrome

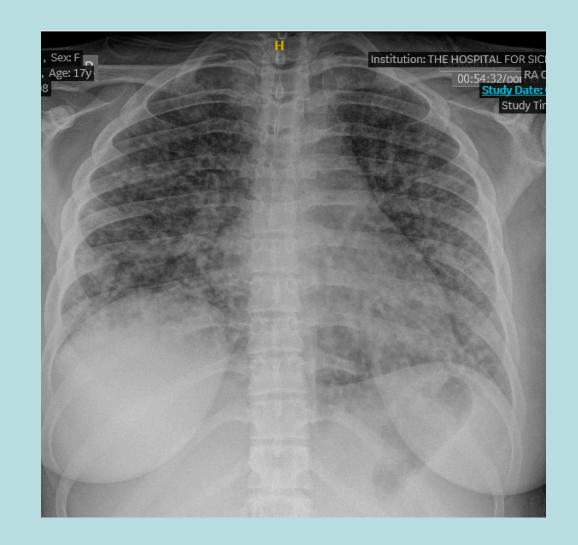
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Case Background

- 17-year-old female born in Pakistan, immigrated to Canada >10y ago
- Presented in November 2021 with 2 months of nausea, vomiting, bloody diarrhea, weight loss, and weakness
 - Colonoscopy revealed moderate to severe chronic active colitis with foci of erosions → Diagnosed with ulcerative colitis
- TST done prior to initiation of biologics was negative
- Started on infliximab
- Two months later, presented with daily fevers, myalgias, nausea and vomiting, weight loss, decreased appetite
 - Productive cough without hemoptysis, dyspnea, orthopnea
- Bronchoalveolar lavage specimens were 2+ acid-fast bacilli and positive on PCR for TB



Initial Decompensation

- Within 45 minutes of initial RIPE therapy, developed hemodynamic instability
 - Hypotension, new oxygen requirements, fever
- Mild splenomegaly on MRI
- Bone marrow aspirate showed hemophagocytosis of both erythroid and lymphoid cells by histiocytes and reduce erythroid precursors with dyspoietic features

Lab	Lab Value
Hemoglobin	68 g/L
WBC	$3.4 \times 10^9 / L$
ANC	0.88 x 10 ⁹ /L
Platelets	31 x 109 /L
ESR	>120 mm/hr
CRP	241 mg/L
ALT	39 U/L
AST	80 U/L
LDH	2703 U/L
Ferritin (Peak)	882 ug/L
Fibrinogen (Trough)	1.2 g/L
Triglycerides (Peak)	3.16 mmol/L
Soluble IL-2	1003 U/mL
CXCL9	17,728 pg/mL

Ongoing Challenges

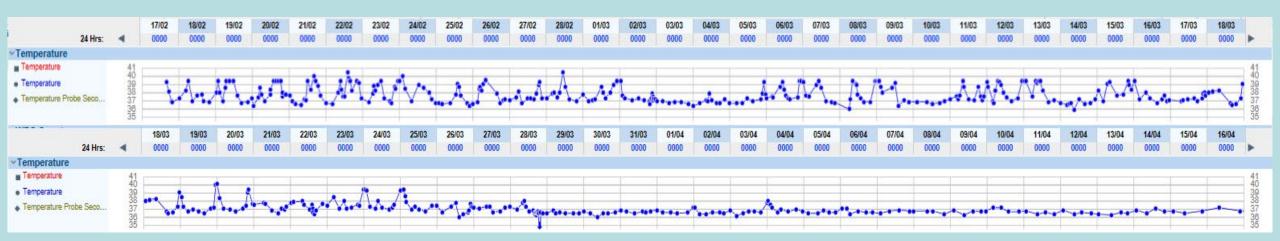
- Fever, nausea, vomiting, anorexia, intolerance of medications
 - Changed to IV moxifloxacin and rifampin
 - Continued enteral isoniazid, pyrazinamide, and ethambuto
- Biochemical decompensation
 - Neutrophil count dropped to 0.12, CRP rose to 250, ferritin rose to 1800
- Anakinra added, attempted steroid wean unsuccessful
 - Neutropenia resolved within 4 days of starting anakinra; other symptoms persisted





Immunosuppression modification

- Anakinra ultimately changed to infliximab to treat TB-IRIS
- Defervesced after first dose of infliximab
- TNF-alpha level normalized from peak of 129pg/mL to 6.3pg/mL
- Anorexia and vomiting ceased
- Steroids tapered successfully





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