Non-cirrhotic Portal Hypertension in Rheumatoid Arthritis

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INTRODUCTION

Gastrointestinal bleeding in rheumatoid arthritis (RA) most commonly occurs from gastroduodenal ulceration in the context of chronic NSAID and prednisone use[1]. We present an unexpected case of variceal bleeding in a patient with RA that was found to be secondary to non-cirrhotic portal hypertension, with liver biopsy revealing nodular regenerative hyperplasia.

CASE PRESENTATION

59-year-old woman with longstanding seropositive rheumatoid arthritis (RA) presented with cough, fevers, hematemesis, and melena.

Medical comorbidities included autoimmune thyroiditis and chronic pancytopenia. Home medications consisted of levothyroxine, chronic prednisone, and ferrous sulfate.

Notably, baseline cytopenia included hemobglobin 9 g/dL, white blood cell count 2 x10^9/L, and platelets 100 x10^9/L.

For the past two years, she had experienced intermittent black tarry stools and abdominal pain associated with fatigue and drenching night sweats.

No source of bleeding was previously found by colonoscopy and upper endoscopy.

In the three weeks leading to hospital admission, she began to experience persistent flu-like symptoms with subjective fever, cough productive of greenish-brown sputum, and black stools. The morning of presentation, she experienced an episode of large volume hematemesis

WORK-UP

- On presentation, she was febrile to 39.4ºC, tachycardic with heart rates up to 130 beats per minute, with blood pressures 101/50 mmHg and respiratory rate of 18 breaths per minute.
- Physician exam notable for BMI 19.5 kg/m2, inspiratory bibasilar crackles were noted on lung exam, right-sided abdominal tenderness with no apparent visceromegaly, subcutaneous rheumatoid nodules on her arms and forearms, and several spider nevi on the upper chest.
- Laboratory evaluation on presentation was notable for the following:
  - WBC 16.8 x10^9/L | Hb 4.7 g/dL | Plt 131 x10^9/L
  - BUN 22 mg/dL | Cr 0.3 mg/dL
  - ESR 160 mm/h | CRP 53 mg/L
  - Total bilirubin 0.5 mg/dL | ALT 62 U/L | AST 131 U/L | Alkaline Phosphatase 296 U/L
- CT Abdomen and Pelvis with contrast revealed bilateral lung interstitial infiltrates, and splenomegaly
- Upper endoscopy was performed, which revealed bleeding esophageal varices, which were subsequently banded.
- Transjugular liver biopsy with hepatic venous pressure gradient (HVPG) was then performed, revealing nodular regenerative hyperplasia (NRH), and HVPG confirmed presence of portal hypertension
- Due to the presence of RA, baseline neutropenia, and splenomegaly, with NRH on liver biopsy, she was diagnosed with Felty Syndrome.

DISCUSSION

- RA often presents with extra-articular features. Felty Syndrome (FS) is a rare extra-articular manifestation of longstanding, joint-deforming, seropositive RA occurring in less than 1% of patients, and is a clinical diagnosis made with the presence of RA, neutropenia, and splenomegaly[2].
- Though not all three features must be present for a diagnosis of Felty syndrome, neutropenia with absolute neutrophil count less than 2000/microL is required for diagnosis. However, the degree of neutropenia may vary depending on clinical course of illness and may even be elevated in acute infection, as in this case[2].
- NRH is portal venopathy that has multiple potential causes including hematologic processes, drug-induced etiologies, and rheumatologic diseases. As endothelial damage occurs, obliterative changes in the portal venous system occur with compensatory hypertrophy and nodule formation, causing surrounding areas to increase blood supply forming NRH[3].
- A strong association exists between Felty Syndrome and NRH. NRH leads to a form of presinusoidal non-cirrhotic portal hypertension, which may lead to variceal formation and bleeding.

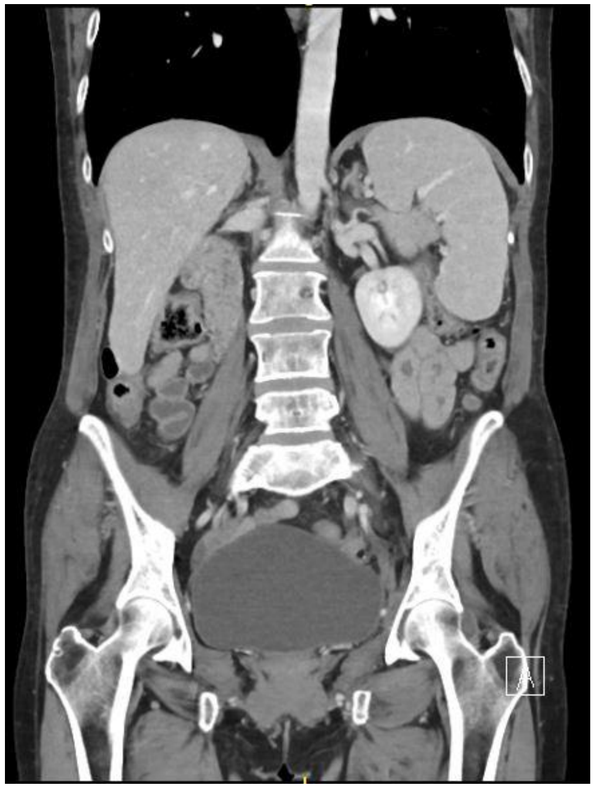

Figure 1: Admission CT scan demonstrating splenomegaly measuring 14 cm

CONCLUSIONS

This case highlights a case of Felty syndrome and emphasizes the importance of maintaining a broad differential to GI bleeding in a patient with rheumatoid arthritis and considering NRH in a patient with long-standing RA that presents with manifestations of portal hypertension.

REFERENCES

1. Myasoedova E, Talley NJ, Manek NJ, Crowson CS. Prevalence and risk factors of gastrointestinal disorders in patients with rheumatoid arthritis: results from a population-based survey in olmsted county, Minnesota. Gastroenterol Res Pract. 2011;2011:745829.
2. Balint GP, Balint PV. Felty's syndrome. Best Pract Res Clin Rheumatol. 2004;18(5):631-645.
3. Patel SM, Tester GA, Munson GW, Sanchez W, Rosen CB, Vege SS. Under Pressure: Where's the Cirrhosis? The American Journal of Medicine. 2011;124(9):818-820.