

Triple Autoantibody Positivity in Isolated Portal Vein Thrombosis: Expanding the Spectrum of Autoimmune Vasculitic Overlap Syndrome

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ABSTRACT:

Background: Autoimmune-mediated vascular injury is an uncommon but important cause of non-cirrhotic portal hypertension (NCPH) and splanchnic vein thrombosis. The concurrent presence of C-ANCA, P-ANCA, and Anti-La (SSB) antibodies has been very rarely reported in association with portal vein thrombosis (PVT).

Case Presentation:

We report a rare case of a 55-year-old female with a known history of ascites for 2 years, presenting with worsening abdominal distension and malena for 15 days. Evaluation revealed non-cirrhotic portal hypertension with thrombosis extending from the portal vein to the confluence and right main portal vein. She tested positive for C-ANCA, P-ANCA, and Anti-La antibodies without systemic vasculitis or classic autoimmune disease features.

Conclusion:

This rare autoimmune overlap syndrome highlights the need to consider vasculitic and autoimmune mechanisms in unexplained PVT in non-cirrhotic patients. Early recognition and immunological testing can guide management and prevent complications.

Keywords: Autoimmune vasculitis, portal vein thrombosis, non-cirrhotic portal hypertension, C-ANCA, P-ANCA, Anti-La, overlap syndrome

INTRODUCTION:

Portal vein thrombosis (PVT) is a rare but serious condition that may arise in both cirrhotic and non-cirrhotic settings, with an incidence of 1% in autopsy studies and 2-4 per 100,000 annually in the general population¹. Cirrhosis and malignancy remain the most common causes; however, 20–25% of PVT cases are idiopathic, prompting evaluation for thrombophilic, infectious, and autoimmune conditions².

Non-cirrhotic portal hypertension (NCPH) results from pre-sinusoidal obstruction, typically due to thrombosis, obliterative portal venopathy, or nodular regenerative hyperplasia. While the pathophysiology often involves increased intrahepatic resistance and collateral formation, autoimmune mechanisms may also contribute^{3,4}.

ANCA-associated vasculitides (AAVs), including granulomatosis with polyangiitis and microscopic

polyangiitis, generally involve small vessels of the lungs and kidneys. Gastrointestinal or portal vasculature involvement is rare and typically occurs late in the disease course⁵. The simultaneous presence of C-ANCA, P-ANCA, and Anti-La antibodies in a patient with portal vein thrombosis and no systemic autoimmune features is extremely rare and suggests an evolving overlap syndrome with vasculitic and autoimmune components.

This report describes such a case, with the goal of expanding differential considerations in idiopathic PVT and highlighting the potential for silent autoimmune vascular injury.

Case Report:

Patient Profile:

A 55-year-old female, known case of chronic ascites for 2 years, presented with complaints of black

coloured stools and progressive abdominal distension for 15 days which was associated with pain in umbilical region and not associated with any aggregating or relieving factors, no diurnal variation was noted. Patient also complains of hematemesis, 20 days back around 3 episode and was about 30 ml in quantity. There was no history of fever, joint pain, rash, dry eyes/mouth, cough, or hematuria. Patient has no previous history of drug intake and was found to be non alcoholic.

Clinical Examination:

- Vitals: Stable
- General physical examination: Pallor and Oedema present
- Abdomen: Moderate ascites, no hepatosplenomegaly
- No signs of chronic liver disease
- No neurological or systemic signs of vasculitis

Laboratory Investigations:

- Hemoglobin: 7.8 g/dL, RBC: Low (2.74)
- WBC/Platelet: Normal
- LFTs: Normal SGOT(14) and SGPT(20), bilirubin; Normal(0.263), ALP: Normal (57 IU/L), Serum albumin: Low(2.7 g/dL)
- PT/INR: Normal (14.4/1.07)
- Viral hepatitis serology and HIV : Negative
- ANA: Negative
- C-ANCA: Positive (PR3) by ELISA (29.08)
- P-ANCA: Positive (MPO) by ELISA (23.64)
- Anti-La (SSB): Positive (9.32)

- Anti-Ro (SSA), dsDNA, anti-Smith, anti scl-70 and antiphospholipid antibodies: Negative
- Serum protein electrophoresis: Normal
- Thrombophilia screen: Negative (Protein C, S, AT-III, Factor V Leiden)

Imaging:

•Ultrasound abdomen with Doppler: (Fig.1)

Liver appears normal in size, shape and shows altered echo texture

Portal vein appears dilated (diameter approx 18mm) with evidence of echogenic material in main portal vein extending upto confluence and right main portal vein s/o thrombus.

Spleen is normal in shape & echo pattern appears mildly enlarged in size splenic vein appears dilated (diameter approx 12.4mm) with multiple collaterals are seen in splenic hilum and porta region.

•CT Abdomen with contrast: Non-cirrhotic liver morphology, portal hypertension, collaterals seen.

•Liver elastography: Liver stiffness not consistent with cirrhosis (4.8 kPa)

Ascitic Fluid Analysis:

- SAAG > 1.1 g/dL, Protein- Low (0.76)
- Total WBC: 30 cells/mm³ (lymphocytic)
- ADA: Normal
- Culture and cytology: Negative

Upper GI Endoscopy:

(Fig.2)- Portal hypertension- oesophageal varices grade 3, 2 columns, RCS+, EVL done and 3 bands applied.

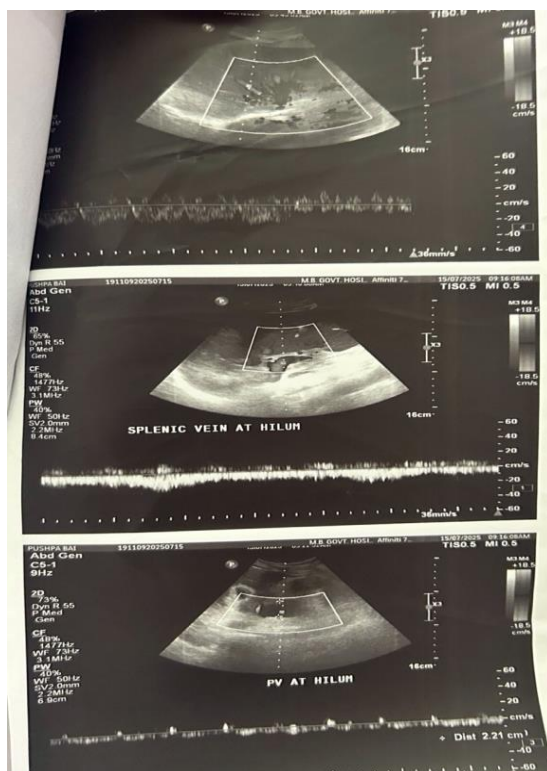


Fig.1: Colour doppler of spleno-portal axis showing portal vein thrombus.

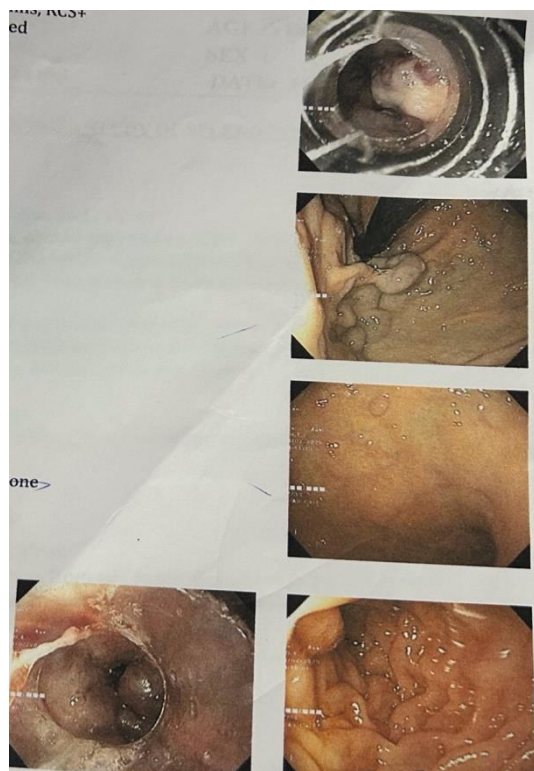


Fig.2 –upper GI Endoscopy showing showing Oesophageal Varices.

Diagnosis:

Autoimmune vasculitic overlap syndrome with isolated extensive portal vein thrombosis and non-cirrhotic portal hypertension

Management and Outcome:

- Anticoagulation with Inj enoxaparin, transitioned to rivaroxaban
- Oral corticosteroids (prednisolone 0.5 mg/kg/day) initiated
- No immunosuppressive agents added at initial stage
- Close follow-up planned for emergence of systemic autoimmune features
- Serial Doppler to monitor clot resolution
- Upper GI Endoscopy showed Grade 3 varices – managed with Endoscopic variceal ligation and bands.

DISCUSSION:

This case illustrates an unusual autoimmune presentation involving splanchnic vasculature in the absence of systemic disease. Simultaneous positivity for C-ANCA and P-ANCA is rare, and when accompanied by Anti-La antibodies, may suggest evolving or silent autoimmune overlap^{5,6}.

Only a handful of case reports describe ANCA-associated vasculitis presenting with gastrointestinal thrombotic manifestations. A recent systematic review of ANCA vasculitis found GI involvement in <5% of cases, with bleeding, ischemia, or infarction more commonly reported than thrombosis⁷.

The mechanism likely involves immune-mediated endothelial injury, leading to local thrombosis and vascular remodelling. Anti-La positivity without sicca symptoms may precede full-blown Sjögren's syndrome or SLE. This aligns with newer findings suggesting a prodromal autoimmune phase in up to 30% of patients with single antibody positivity⁸. Given the extent of thrombosis and absence of cirrhosis, early anticoagulation was appropriate. In selected autoimmune PVT cases, steroids or additional immunosuppressants may be beneficial if systemic features develop^{9,10}. This report supports the inclusion of extended autoimmune work-up in patients with idiopathic PVT and NCPH, especially when thrombotic extent is disproportionate and liver function is preserved.

CONCLUSION:

Triple positivity for C-ANCA, P-ANCA, and Anti-La antibodies in a non-cirrhotic patient presenting solely with portal vein thrombosis and ascites represents a highly unusual autoimmune vasculitic overlap. This case underlines the diagnostic value of immunological testing in unexplained PVT and expands the phenotype of autoimmune vascular disease beyond classical organ involvement.

Patient Consent:

Informed written consent was obtained from the patient for publication.

Conflict of Interest:

The authors declare no conflicts of interest.

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