

Abstract

Polyarteritis Nodosa is a medium vessel vasculitis that has multiple organ involvement. Most common GI tract manifestations include mesenteric ischemia however rarely it can involve splenic vasculature leading to splenic infarct. Its pathological manifestations are limited to the arterial system favoring branch point of arteries and may involve celiac trunk and its distal branches. Early detection is key in preventing serious complications and progression of disease. Medical therapeutics such as steroids and cyclophosphamide are indicated for severe disease that does not qualify for vascular or surgical intervention. Follow up imaging is indicated to monitor for treatment response and disease progression.

Background

Polyarteritis Nodosa (PAN) most typically affects medium and small sized vessels. It usually presents with non-specific symptoms of fatigue, myalgia and arthralgias; however, vasculitis is a late presentation. As it commonly involves vital organs such as GI tract and kidneys, delay in treatment can have a high mortality rate due to potential ischemia or micro aneurysmal rupture.

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Gastrointestinal Manifestations of Polyarteritis Nodosa

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Case

A 50 year old female with history of smoking presented with sharp abdominal pain radiating to left upper quadrant and back. She denied any associated nausea/vomiting/diarrhea or constipation but reported joint pain in toes and fingers, and nodules in her pubic area.

Physical exam was remarkable for palpable nontender raised skin nodule on suprapubic region and bilateral palmar erythema.

Laboratory analysis was significant for elevated rheumatoid factor at 78, mildly elevated ESR 36 and CRP 13.8, complement C3 C4 within normal limits, negative Hepatitis B, C, and negative ANCA and ANA.

A CT of abdomen and pelvis revealed a 7.2 x 2.9 x 6.6 cm splenic infarct, perivascular infiltration in the celiac artery extending to common hepatic artery and short gastric artery (as shown in Image 1).

Due to high suspicion of vasculitis, the patient underwent biopsy of the suprapubic nodule, which showed a hemorrhagic nodule with organizing abscess.

Imaging



Image 1. Ring of low attenuation seen around celiac artery extending to involve common hepatic, short gastric and splenic artery (shown with the arrow)

Clinical Course

The distribution of vessel involvement along the branches of celiac artery on imaging in addition to the biopsy results suggested PAN. The patient received pulse dose steroids with IV 250mg methylprednisone for 3 days. She was discharged on prednisone taper and started on Cyclophosphamide therapy outpatient which resulted in symptomatic improvement.

References

1. Goldman L, Schafer A. Goldman-Cecil Medicine, 26th ed. New York, NY. Elsevler Inc; 2020.

2. Hekali P, Kajander H, Pajari R, Stenman S, Somer T. Diagnostic Significance of Angiographically Observed Visceral Aneurysms with Regard to Polyarteritis Nodosa. Acta Radiologica. 1991;32(2):143-148. doi:10.1177/028418519103200212 3. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Polyarteritis Nodosa. Arthritis Care and Research, 2021; 73 (8) 1061-1070. Doi: 10.1002/acr.24633 4. Unverdi S, Altay M, Duranay M, Krbas I, Demirci S, Yuksel E. Polyarteritis Nodosa Presenting with Splenic Infarction, Bilateral Renal Infarction, and Hematoma. Southern Medical Journal. 2009; 102 (9): 972-973. doi: 10.1097/SMJ.0b013e3181b21a29.

5. Lin, Y.C., Kalot, M.A., Husainat, N.M., Byram, K., Dua, A.B., James, K.E., Springer, J.M., Turgunbaev, M., Villa-Forte, A., Abril, A., Langford, C., Maz, M., Chung, S.A. and Mustafa, R.A. (2021), Polyarteritis Nodosa: A Systematic Review of Test Accuracy and Benefits and Harms of Common Treatments. ACR Open Rheumatology, 3: 91-100.



Discussion

PAN has a predilection for skin, peripheral nerves, the GI tract and the kidneys. A classic GI manifestation is mesenteric vasculitis leading to transmural necrotizing inflammation and bowel ischemia¹. The pathological changes of PAN are limited to the arterial system favoring branch point and thus can be detected with CT or angiography, which is a gold standard for assessment with a sensitivity of 89% and a specificity of 90% ^{2,3}.

Our case demonstrates a rare complication of PAN associated vasculitis. Vascular imaging demonstrated celiac trunk involvement that extended into splenic artery leading to subcapsular splenic infarction whereas mesenteric, common hepatic and bilateral renal arteries appeared patent. Timely diagnosis of this condition is important to prevent progression which can include splenic rupture.⁴

Our patient experienced symptomatic improvement after guideline recommended medical therapeutics were initiated with pulse dose steroids and cyclophosphamide.⁵ Additionally, follow up abdominal vascular imaging is recommended to assist with assessment of disease control and treatment response even in patients who become clinically asymptomatic.³