CASE REPORT

Polyarteritis nodosa presenting atypically as appendicitis and pyelonephritis in a single patient

Matthew Corbitt,¹ Jagadeesh Kurtkoti,^{2,3} Muddassir Rashid,⁴ Sonu Nigam^{1,5}¹Griffith University School of Medicine, Gold Coast, Queensland, Australia²School of Medicine, Griffith University – Gold Coast Campus, Southport, Queensland, Australia³Department of Renal Medicine, Gold Coast University Hospital, Southport, Queensland, Australia⁴Department of Radiology, Gold Coast University Hospital, Southport, Queensland, Australia⁵Department of Pathology, Gold Coast University Hospital, Southport, Queensland, Australia**Correspondence to**Dr Jagadeesh Kurtkoti,
Jagadeesh.Kurtkoti@health.qld.gov.au

Accepted 22 December 2018

SUMMARY

A 17-year-old man presented to the emergency department 10 days postlaparoscopic appendectomy with ongoing lower abdominal pain. The pain was associated with fevers, chills, sweating and constipation. There were no other associated symptoms. On examination, there was suprapubic and right lower quadrant tenderness and an elevated blood pressure (140/90 mm Hg). The patient later developed severe bilateral flank pain. Investigations revealed elevated C reactive protein, leukocyturia and microscopic haematuria. Blood and urine cultures were negative. CT angiogram demonstrated bilateral wedge-shaped peripheral renal hypodensities suggestive of several peripheral infarcts with intrarenal microaneurysms. Treatment with steroids and cyclophosphamide was initiated, leading to significant clinical improvement. Review of the histological appendix specimen revealed features consistent with small-medium vessel vasculitis.

BACKGROUND

This case demonstrates two atypical presentations of polyarteritis nodosa (PAN) in a single patient, presenting first as appendicitis and then pyelonephritis.

CASE PRESENTATION

A 17-year-old man presented to the emergency department 10 days postlaparoscopic appendectomy for uncomplicated appendicitis with ongoing lower abdominal pain, most prominent in the right iliac fossa. The pain was associated with fevers, chills, night sweats and constipation. He denied nausea, vomiting, shoulder tip pain, dysuria. His vitals were only remarkable for an elevated blood pressure (140/90 mm Hg).

On examination, he was well perfused with no signs of peritonitis. There was suprapubic and right lower quadrant tenderness. There was no abdominal distension and the surgical sites were unremarkable.

Investigations showed only an elevated white cell count ($16.0 \times 10^9/L$) and elevated C reactive protein (217 mg/L). A CT-abdomen revealed bilateral wedge-shaped renal hypodensities and a provisional diagnosis of pyelonephritis was made. The patient was started on intravenous antibiotics.

After 5 days, the patient's condition had not improved, and he had subsequently developed severe bilateral flank pain and intermittent abdominal cramping. Vitals were significant for an elevated

blood pressure (164/92 mm Hg) and fever. Further investigations revealed a continued elevation of C reactive protein (235 mg/L), white cell count ($14.8 \times 10^9/L$) and new elevations in lactate dehydrogenase (460 u/L) and creatinine (94 $\mu\text{mol/L}$, baseline 76 $\mu\text{mol/L}$). Urine microscopy culture sensitivities showed leukocyturia and microscopic haematuria but no bacterial growth. Antinuclear antibody, antineutrophil cytoplasmic antibody, lupus antibody, Hepatitis B and C serology, thrombophilia screen and sexually transmitted infection screen were negative. Multiple blood cultures were negative.

An abdominal CT-angiogram was performed and demonstrated bilateral wedge-shaped peripheral renal hypodensities suggestive of several peripheral infarcts with intrarenal microaneurysms (figure 1). Further questioning revealed that the patient had a 3-month history of subjective fevers, 20 kg weight loss and bilateral wrist and elbow arthralgia without swelling. A diagnosis of PAN was made and the patient was started on a course of high-dose of intravenous steroids and cyclophosphamide, which led to substantial clinical improvement.

Review of the histological appendix specimen (figure 2) was undertaken and demonstrated small to medium-sized vessel vessels showing fibrinoid necrosis and mixed inflammatory infiltrate comprised of lymphocytes, eosinophils and histiocytes. There was also endothelial swelling and red blood cell extravasation. These histological features are consistent with small-medium vessel vasculitis.

INVESTIGATIONS

As described above.

DIFFERENTIAL DIAGNOSIS

As described above.

TREATMENT

As described above.

OUTCOME AND FOLLOW-UP

The patient was discharged on prednisolone, pantoprazole, cyclophosphamide, trimethoprim/sulfamethoxazole and calcitriol and advised to have regular follow-up as an outpatient in the renal clinic. The patient has remained well since discharge.

DISCUSSION

PAN is a systemic necrotising vasculitis that typically affects small-to-medium size arteries. It can

© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Corbitt M, Kurtkoti J, Rashid M, et al. *BMJ Case Rep* 2019;**12**:e228072. doi:10.1136/bcr-2018-228072

Figure 1 (A) CT-abdomen demonstrating multiple peripheral wedge-shaped hypodensities (green arrows) in keeping with infarcts, and numerous microaneurysms arising from the intralobar arteries bilaterally (blue arrows). (B) 3D reconstruction demonstrating numerous microaneurysms bilaterally (blue arrows).

manifest as either single organ or multiorgan disease, most often affecting peripheral nerves and skin,¹ but sparing the lungs.² Renal, cardiac, gastrointestinal tract and neurological system involvement is also seen, making the presentation of PAN quite variable. Its epidemiology is difficult to determine; however, it affects men more frequently than women and the average age of onset is 50 years.² PAN can be classified into three subtypes: idiopathic generalised PAN, cutaneous PAN and hepatitis B virus-associated PAN.²

PAN typically presents with constitutional symptoms that are often only recognised retrospectively. These include weight loss, night sweats, malaise, fevers and myalgias. Renal manifestations include infarcts and hypertension secondary to intrarenal artery involvement. Gastrointestinal tract involvement is often serious and presents in one-third of cases as a surgical abdomen. The most common neurological finding is mononeuritis multiplex and cutaneous features include purpura, subcutaneous nodules,

Figure 2 (A) H&E x100. Appendix showing necrotising vasculitis (blue arrow). (B) H&E x400. Medium sized vessel showing fibrinoid necrosis (yellow arrow) and transmural inflammatory infiltrate comprised of eosinophils (black arrow), histiocytes and lymphocytes.

necrotic ulcers and livedo reticularis. Interestingly, the most characteristic feature of PAN is orchitis, secondary to testicular artery ischaemia, although this is rarely seen clinically.²

Diagnosing PAN is challenging, as it is not possible to make a diagnosis through clinical examination or serological testing alone, and atypical presentations occur.³⁻⁵ Diagnosis is dependent on a combination of clinical, radiological and histological findings. Suspicion is usually raised when a patient has constitutional symptoms with multisystem involvement.²

Treatment for PAN where a major organ or body system is involved initially requires high-dose glucocorticoids plus

Learning points

- ▶ Polyarteritis nodosa (PAN) is a small-medium vessel vasculitis that can manifest as single or multiorgan disease, sparing the lungs.
- ▶ PAN can present atypically and be missed as appendicitis or pyelonephritis.
- ▶ Diagnosis of PAN requires judicious appraisal of a patient's clinical history, examination findings, radiological and histological findings, with the gold standard being biopsy.

intravenous cyclophosphamide. For remission maintenance therapy, low-dose glucocorticoids and azathioprine are used.⁶

This case demonstrates other atypical presentations of PAN, presenting as both appendicitis and pyelonephritis in a young man without cutaneous, neurological or recognisable serological features.

Contributors MC wrote the manuscript. JK reviewed and submitted the manuscript. MR reviewed images. SN reviewed pathology images.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

REFERENCES

- 1 Hernández-Rodríguez J, Alba MA, Prieto-González S, *et al.* Diagnosis and classification of polyarteritis nodosa. *J Autoimmun* 2014;48-49:84–9.
- 2 De Virgilio A, Greco A, Magliulo G, *et al.* Polyarteritis nodosa: A contemporary overview. *Autoimmun Rev* 2016;15:564–70.
- 3 Andrade CA, Vasconcelos A, Pinto JC. Polyarteritis nodosa: an atypical presentation. *Case Rep* 2015;2015:bcr2015209564.
- 4 Shumy F, Anam AM, Chowdhury MAJ. Spontaneous bilateral perirenal and splenic haematoma in childhood onset polyarteritis nodosa. *BMJ Case Rep* 2018;2018:bcr-2018-225762.
- 5 Starobinska E, Sekhon K, Bilal J. Abdominal pain leading to incidental finding of polyarteritis nodosa. *BMJ Case Rep* 2018;2018:bcr-2018-225438.
- 6 Mukhtyar C, Guillevin L, Cid MC, *et al.* EULAR recommendations for the management of primary small and medium vessel vasculitis. *Ann Rheum Dis* 2009;68:310–7.

Copyright 2019 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow