CASE REPORT

Spontaneous bilateral perirenal and splenic haematoma in childhood onset polyarteritis nodosa

Farzana Shumy,¹ Ahmad Mursel Anam,² Md Abdul Jalil Chowdhury¹

¹Department of Internal Medicine, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

²Intensive Care Unit, Square Hospitals Ltd, Dhaka, Bangladesh

Correspondence to
Dr Ahmad Mursel Anam,
murselanam@gmail.com

Accepted 12 June 2018

SUMMARY

Spontaneous bilateral perirenal haematoma, as well as splenic haematoma, are rare occasions in polyarteritis nodosa (PAN). We report a case of a young man, who suffered from various typical and atypical manifestations of PAN since childhood. The diagnosis was delayed due to symptoms mimicking with other clinical conditions. Finally, the diagnosis was confirmed when presented with perirenal and splenic haematoma and was managed successfully.

BACKGROUND

Polyarteritis nodosa (PAN) is a systemic necrotising vasculitis of small-sized and medium-sized arteries. Among various presentations, spontaneous perinephric haematoma (SPH) is a rare, as well as, catastrophic presentation. Moreover, splenic haematoma is also an infrequent complication of PAN.^{1,2} A young man suffered a lot for a long time, because the diagnosis was missed in each presentation due to mimicking with other illness. Vasculitis has diverse manifestations, and we should be aware of both typical and atypical presentations. We believe that the journey of the patient could be an eye-opener to clinicians, and it will remind the importance of detailed history taking and correlation, as well. Timely diagnosis saved the life of the patient and enabled him to lead a further uneventful life.

CASE PRESENTATION

In April 2015, an 18-year-old man was admitted in a tertiary care hospital for evaluation of his recurrent severe abdominal pain for the past 3 months.

He had his first sufferings in mid-January, 2015. At that time, he was hospitalised and a CT scan of abdomen revealed splenomegaly with features of intrasplenic haemangioma and left perinephric haematoma. There was no history of trauma or other obvious reason for the intra-abdominal bleed. He was managed conservatively and discharged with alleviation of pain only. Further evaluation was not done due to unwillingness of patient's attendant's to stay in hospital. Second episode of abdominal pain was two-and-half months later, when he developed shock. He was managed with transfusion of packed cell. This time another CT scan of abdomen revealed large right posterior-inferior perinephric collection, multiple small splenic haematoma and small left perinephric collection [figure 1](#). During this period of illness, he lost about 11 kg weight. He became bedbound due to severe weakness and lethargy and continued to suffer from on-and-off abdominal pain. We found him extremely ill looking with raised blood pressure (140/100 mm Hg) and raised temperature (~38°C). Abdomen examination showed splenomegaly and palpable right para-umbilical mass.

We reviewed his history and found that his illness actually started long ago. At the age of 8 years, he developed arthralgia and on-and-off low grade fever, malaise and anorexia. Since then he was on NSAID as-per-need basis. At the age of 9, he had multiple painful nodular skin lesions that healed spontaneously. At the age of 10, due to severe myalgia, he was treated with prednisolone, presumed to be a case of viral myositis and recovered completely. In 2007, when he was 11-year-old, he was treated with methotrexate and naproxen for his multiple joint pains, being diagnosed as juvenile rheumatoid arthritis. He was partially improved with occasional exacerbation of joint pains and fever. Since then he was on methotrexate and on-and-off prednisolone. In 2009, he was hospitalised with acute abdomen and underwent laparotomy and repair of perforated ileum, and he was further managed as a patient of enteric fever.

In 2011, prednisolone was discontinued as he developed hypertension. About 9 months following discontinuation of prednisolone, he was again hospitalised due to headache, double vision, gait abnormalities and tremor. Documents showed that he had multiple cranial nerve palsy and cerebellar signs, and once more received prednisolone. MRI of brain was normal and cerebrospinal fluid was negative for oligoclonal bands and his neurological features improved with steroid.

Figure 1 CT scan of the abdomen showing large right posterior-inferior perinephric collection, multiple small splenic haematoma and small left perinephric collection.

To cite: Shumy F, Anam AM, Chowdhury MAJ. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2018-225762

Figure 2 DSA showing two small aneurysms in the lower pole of the left kidney. DSA, digital subtraction angiography.

Figure 3 DSA showing two moderate size aneurysms in the splenic artery. DSA, digital subtraction angiography.

INVESTIGATIONS

This time, blood investigations were unremarkable except low haemoglobin (10.5 g/dL) and raised erythrocyte sedimentation rate (62 mm in first hour) and C reactive protein (158 mg/dL). Coagulation parameters were also normal. Urinalysis showed pus cell 0–2/high power field (HPF) and RBC 7–10/HPF (5% dysmorphic) with normal renal function. Antinuclear antibodies, antineutrophil cytoplasmic antibodies and serology for hepatitis B and C were negative. Subsequently, he underwent digital subtraction angiography. It revealed two small aneurysms in lower poles of both kidneys [figure 2](#), a small aneurysm in each of the inferior mesenteric and left gastric artery, and two moderate size aneurysms in the splenic artery [figure 3](#).

TREATMENT

He was started on intravenous methylprednisolone followed by oral prednisolone and pulse cyclophosphamide (750 mg/m² body surface area) continued for 12 months along with antihypertensive medication. He was switched to azathioprine along with low-dose steroids.

OUTCOME AND FOLLOW-UP

He was completely symptoms free at 12 months of follow-up. He remained asymptomatic with no recurrence of symptoms on further follow-up.

DISCUSSION

Spontaneous perinephric haemorrhage (Wunderlich syndrome) is an uncommon condition. Moreover, secondary to PAN is rarely seen. Zhang *et al*³ reported that SPH was associated with tumour (61.5%), vascular disease (17%) and infection (2.4%). In 1908, Schmidt⁴ first described perirenal haematoma by ruptured of microaneurysm in PAN. In a meta-analysis, only five cases of perirenal haematoma due to PAN has been described.^{1 5 6}

Management of ruptured aneurysm resulting perirenal haematoma is difficult. In acute setting, nephrectomy is associated with high mortality rate. Sometimes urgent nephrectomy is lifesaving. Few cases need conservative drainage and sometimes selective arterial embolisation is also necessary in cases of active bleeding. However, reduction or resolution of aneurysm is also possible, due to resolution of inflammation of vessels wall.^{1 7}

PAN is a necrotising inflammation of medium-sized or small-sized arteries. It typically develops subacutely with the onset of constitutional symptoms. Commonly described clinical features are livedo reticularis, nodules, mononeuritis multiplex, abdominal pain and testicular involvement. Regarding neurological involvement, peripheral nervous system involvement is more frequent than central nervous system. Previously reported cases of neurological involvement have included encephalitis, seizures, hemiparesis, psychosis, subarachnoid haemorrhage and rarely cranial nerve palsy.^{7 8 9}

Renal involvement in PAN has been described in the form of microaneurysms, multiple renal infarcts, proteinuria, hypertension and renal insufficiency. Involvement of splenic arteries and splenic haematoma is also a rare occasion in PAN. Sigmoid perforation and mesocolic haematoma have also been reported.⁷

Our patient presented with various typical and atypical features of PAN. His clinical features covered almost all the segment of rare features of PAN. Unfortunately diagnosis was delayed because of sequential presentation of sign and symptoms, mimicking other diseases. Moreover, childhood PAN is also a rare occasion. Careful review and meticulous history might help physician to establish early diagnosis. However, treatment

was started during life-threatening situation and successfully managed only with immunosuppressive medications with no recurrence of symptoms.

Learning points

- ▶ History taking is an important issue for every clinician. This case reminds us that proper history taking is always essential for correct diagnosis.
- ▶ Management with only immunosuppressive drugs is possible in polyarteritis nodosa (PAN) presented with perinephric haematoma.
- ▶ Childhood onset PAN remains challenging for diagnosis and management.

Contributors FS and AMA: contributed to the conception, design, analysis and interpretation of the data; responsible for the drafting the article and the final approval of the version to be published; revising the manuscript critically for important intellectual content. FS and MAJC: contributed in treatment of the patient. All authors: read and approved the final manuscript.

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.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Venkatramani V, Banerji JS. Spontaneous perinephric hemorrhage (Wunderlich syndrome) secondary to polyarteritis nodosa: Computed tomography and angiographic findings. *Indian J Urol* 2014;30:452–3.
- 2 Unverdi S, Altay M, Duranay M, *et al.* Polyarteritis nodosa presenting with splenic infarction, bilateral renal infarction, and hematoma. *South Med J* 2009;102:972–3.
- 3 Zhang JQ, Fielding JR, Zou KH. Etiology of spontaneous perirenal hemorrhage: a meta-analysis. *J Urol* 2002;167:1593–6.
- 4 Schmidt JE. Uber periarteritis nodosa. *Beitr Pathol Anat* 1908;43:455–9.
- 5 Choi HI, Kim YG, Kim SY, Jeon DW, *et al.* Bilateral Spontaneous Perirenal Hemorrhage due to Initial Presentation of Polyarteritis Nodosa. *Case Rep Med* 2015;2015:1–4.
- 6 Oksüzöglü BC, Oksüzöglü G, Tosun E, *et al.* Perirenal haematoma as the presenting feature of polyarteritis nodosa: is it more common in Turkey? *Nephrol Dial Transplant* 1997;12:582–4.
- 7 Mukhopadhyay P, Rath M, Kohli HS, *et al.* Polyarteritis nodosa presenting with spontaneous perirenal hematoma. *Indian J Nephrol* 2012;22:295–7.
- 8 Guirola R, Hunter JV, Perez M, *et al.* Childhood polyarteritis nodosa presenting with central nervous system manifestations and the posterior reversible encephalopathy syndrome. *J Child Neurol* 2014;29:103–7.
- 9 Wahezi DM, Gomes WA, Ilowite NT. Cranial nerve involvement with juvenile polyarteritis nodosa: clinical manifestations and treatment. *Pediatrics* 2010;126:e719–e722.

Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-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