CASE REPORT

Case Report: Polyarteritis nodosa or complicated Henoch-Schonlein purpura, a rare case [version 1; peer review: 2 approved with reservations]

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Abstract

Background: Polyarteritis nodosa (PAN) is a vasculitis that affects medium sized arteries. PAN is a rare disease and requires a high vigilance for diagnosis. For instance, PAN and Henoch-Schonlein purpura (HSP) have narrowing differential diagnosis. Here, we report a case of PAN.

Case presentation: Our patient was a 65 year old woman that came to hospital due to abdominal pain and skin lesion on the right upper and right lower extremities. All rheumatologic tests were negative. A biopsy of the skin lesion was reported as mild hyperkeratosis, slight spongiosis with intact basal layer. The dermis showed moderate to severe perivascular PMN infiltration with vessel wall degeneration and extravasation of RBCs. A colonoscopy reported diffuse mucosal erythema and erosions were seen in the rectum until 6cm of anal verge. An electromyogram test and nerve conduction velocity study of the upper extremities reported bilateral mild carpal tunnel syndrome, and in the right lower extremities mononeuritis multiplex could not be ruled out. Abdominopelvic CT scan reported diffuse wall thickening of terminal ileum associated with mesenteric fat and narrow enhancement of inferior Mesenteric artery with patchy filling defect. After evaluation, the patient received corticosteroid pulses plus cyclophosphamide.

Conclusion: Diagnosis and treatment of PAN is important and PAN should be considered in a patient with skin lesions and neurological impairment.

Keywords

Polyarteritis nodosa, Henoch-Schonlein purpura, vasculitis

Open Peer Review

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Any reports and responses or comments on the article can be found at the end of the article.

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Competing interests: No competing interests were disclosed.

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Introduction

Polyarteritis nodosa (PAN) is a systemic vasculitis that mostly involves medium sized arteries, and sometimes involves small arteries¹. The prevalence of PAN is estimated to be 2 to 33 million individuals worldwide^{2,3}. The annual incidence in some areas of Europe estimate 4.4 to 9.7 per million population⁴. The diagnosis is most commonly made in middle-aged or older adults, and increases with age, and its peak is in the sixth decade of life². Polyarteritis nodosa can mimic the clinical manifestations of Henoch-Schonlein purpura (HSP). It is difficult to differentiate between PAN and HSP at an early stage. If PAN is not diagnosed and treated at an early stage it has a high morbidity⁵. Considering that PAN is a rare disease and requires a high clinical suspicion for diagnosis, here, we report a case of PAN and the reasoning behind its diagnosis in our patient.

Case report

Patient information

The patient was a 65 year old woman from the south of Iran that came to our hospital due to abdominal pain and skin lesion on right upper and right lower extremities, which were mostly on the distal of extremities since 2 weeks preadmission. Other complaints of the patient were diarrhea, vomiting, chills, fever and anorexia. In the past medical history, the patient had diabetes, hypertension and Bell's palsy (treated with 40mg prednisolone daily).

Clinical findings

On examination of the skin, the patient had palpable plaque in the erythematous and purpuric context with vesicular and bulla lesion on right upper and right lower extremities that mostly extended to the distal part (Figure 1). An abdominal examination revealed mild tenderness in the epigaster. The extremities were warm and end pulses were normal. Neurologic exam of the right lower extremity revealed decreased motor function (muscle power 4/5).

Figure 1. Palpable plaque in the erythematous and purpuric context.

Diagnostic assessment

Laboratory tests: HCV, HBV, HIV, ANA (antinuclear antibodies), cryoglobulin, anti-double-stranded DNA (dsDNA) antibodies, complement (C3 and C4), perinuclear antineutrophil cytoplasmic antibodies (P-ANCA and C-ANCA), all were normal. Urine analysis, amylase and lipase levels were normal. ESR was 40mm/h (normal range, <20mm/h), occult blood one pluses positive, and hemoglobin was 11/9 g/L (normal range, 13–16g/l).

Skin biopsy: Mild hyperkeratosis, slight spongiosis with intact basal layer. The dermis showed moderate to severe perivascular PMN infiltration with vessel wall degeneration and extravasation of RBCs. A diagnosis of a vasculitis leukocytoclastic variant (immunofluorescence is not available at our center).

Evaluation of patient anemia and GI tract were done via endoscopy and colonoscopy.

Endoscopy: Patchy erythematous lesions were observed.

Abdominopelvic CT scan (Figure 2): A 130mm of segment of terminal ileum had diffuse wall thickening (3–8mm) associated with mesenteric fat. Narrow enhancement of inferior mesenteric artery with patchy filling defect, poor enhancement of terminal branches. Therefore, suspicions were: 1)vasculitis, 2)mesenteric ischemia.

Colonoscopy: Diffuse mucosal erythema and erosions were seen in the rectum until 6cm of anal verge. Hemorrhoid without active bleeding in anus, few erythema and ophtus ulcer in cecum. Terminal ileum was not intubated. A diagnosis of a rectal erosion maybe due to vasculitis.

Electromyogram test and nerve conduction velocity: Upper extremities reported bilateral mild carpal tunnel syndrome, and in right lower extremities mononeuritis multiplex could not be ruled out.

Figure 2. Abdominopelvic CT scan with IV contrast. Narrow enhancement of the inferior mesenteric artery can be observed (blue arrow).

Echocardiography: No evidence of any other disorder.

Final diagnosis: Vasculitis (PAN or complicated HSP)

Therapeutic intervention

The patient received 1000 mg methylprednisolone IV pulse daily for 3 days, and 750mg cyclophosphamide IV pulse every two weeks for 3 weeks.

Follow-up and outcomes

After 24 hours of receiving treatment, the symptoms of the patient subsided, and after one week improved skin lesions. Currently, the patient is being treated with 50mg prednisolone daily and then we will taper this amount.

Discussion

Unlike other vasculitis, such as microscopic polyarthritis or Wegener's, PAN is not associated with ANCA⁶. The organs most affected in PAN are the skin, renal and GI tract. Cardiac involvement can manifest itself with hypertension, or even ischemic heart disease⁷. In the skin, PAN may manifest by erythematous nodules, livedo reticularis, ulcer, bullous or vesicular eruption and purpura^{6,8,9}. Gastrointestinal symptoms that may be seen include abdominal pain, nausea, vomiting, melena, and bloody or non-bloody diarrhea¹⁰. One of the most common manifestations of patients with PAN is mononeuropathy multiplex that typically involves both motor and sensory deficits in up to 70% of patients^{6,11}. Most cases of PAN are idiopathic, although hepatitis B virus infection, hepatitis C virus infection, and hairy cell leukemia are important in the pathogenesis of some cases^{3,4,12,13}. PAN can mimic the clinical manifestations of HSP. It is difficult to differentiate between PAN and HSP at an early stage⁵. The biopsy pattern helps to differentiate between PAN and

HSP; in tissue studies of HSP leukocytoclastic vasculitis in post capillary venules together with IgA deposition is observed¹⁴. As already mentioned, PAN is most commonly seen in middle-aged or older adults³, while HSP is a childhood disease that occurs between the ages of 3 and 15 years¹⁵. Neurologic manifestation in HSP is rare. Single reports and case series document neurologic manifestations including headaches, intracerebral hemorrhage, focal neurologic deficits, ataxia, seizures, and central and peripheral neuropathy in children with HSP¹⁶. In the present case, using clinical manifestations and laboratory tests, we excluded other differential diagnosis apart from PAN. Considering that PAN and HSP have narrowing clinical manifestation, we differentiated between the two diseases by age and neuropathy. However, although the diagnosis of the present patient is PAN, for a better diagnosis, immunofluorescence of the biopsy is needed, which is not available in our center. Overall, diagnosis and treatment of PAN is important, and PAN should be considered in a patient with skin lesions and neurological impairment.

Consent

Written informed consent was obtained from the patient for the publication of the patient's clinical details and accompanying images.

Competing interests

No competing interests were disclosed.

Grant information

The author(s) declared that no grants were involved in supporting this work.

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Reviewer Report 13 March 2018

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Patricia Woo

Division of Infection and Immunity, University College London, London, UK

This is indeed a severe case of systemic vasculitis. The criteria used to classify the combined clinical and histological findings are not clearly referenced for discussion. The EULAR/PRINTO/PRES criteria published in 2016 are more discriminatory with the addition of IgA immunofluorescence for IgA associated vasculitis (formerly known as HSP). Unfortunately the authors were not able to perform this test and the skin histology did not describe a necrotizing vasculitis. The authors are correct that mononeuritis multiplex would be a separator, but the evidence in this case is more circumstantial as reported. The time line of Bells palsy is suggestive. The nerve conduction report is equivocal. Renal dysfunction or joint involvement were not mentioned.

Both conditions can respond well to prednisolone alone, or in combination with cyclophosphamide depending on the severity.

Since the case report is to highlight the overlapping features of the vasculitides, I agree that the differentiating diagnostic features are important.

Is the background of the case's history and progression described in sufficient detail?

Partly

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

No

Is the case presented with sufficient detail to be useful for other practitioners?

Partly

Competing Interests: No competing interests were disclosed.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Author Response 16 Mar 2018

Saeid Jokar, Yasouj University of Medical Sciences, Yasouj, Iran

Hi Patricia

I corrected the article in accordance with your comments.

Thank you for advising us on improving the content of the article.

Best Regards

Competing Interests: No competing interests were disclosed

Reviewer Report 25 January 2018

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Marco de Vincentiis

Department of Oral and Maxillofacial Sciences, Sapienza University of Rome, Rome, Italy

The authors present a case of a 65 year old woman that was admitted to the authors' hospital due to abdominal pain and skin lesion on the right upper and right lower extremities, with negative rheumatologic tests. After careful diagnostic workout, the final diagnosis was vasculitis (Polyarteritis nodosa or complicated Henoch-Schonlein purpura). The paper is interesting, also due to the rarity of the disease. However, there are some points that need to be improved before final approval:

1. The authors report Bell's Palsy in patient's medical history; it would be interesting to know when this occurred. Since this was treated with corticosteroid therapy, it is important to know if it was present when the patient already had PAN lesions - and in this case if they also improved during therapy - or if it occurred prior to PAN-related lesions arose.
2. Timing details are missing: in the case presentation - follow up and outcomes - the authors report that the patient "is being treated with 50mg prednisolone daily and then we will taper this amount". It is important to define for how long the patient is being treated, how

much time past from initial diagnosis and therapy, and how the authors intend to reduce treatment.

3. Sensorineural Hearing Loss is often reported in PAN and, in some cases, may occur as the presenting symptom. Hearing Loss is typically bilateral and symmetrical, with sudden or rapidly progressive onset. It would be interesting to know if hearing impairment was reported by the patient, or investigated.
4. In the discussion we recommend to further discuss the findings that led to PAN diagnosis in this case.
5. The paper has several English language typos and grammar mistakes that should be corrected by a native English speaker (i.e. "which were was mostly", etc).

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Is the background of the case's history and progression described in sufficient detail?

Partly

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Partly

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Author Response 29 Jan 2018

Saeid Jokar, Yasouj University of Medical Sciences, Yasouj, Iran

Hi Marco,

Thank you for attention in review of our article. I will correct the article by your statements.

1. One week pre-admission the patient was under corticosteroid treatment because

bell's palsy.

2. After 2 weeks, if the no recurrent of patient symptoms we will taper off corticosteroids amount 10 %. We will reduce the dose of corticosteroids until reaches to control of patient symptoms, then by the patient condition will decide about it.

3. Thank you - I add your statement to the manuscript
Other comments are considered.

Competing Interests: No competing interests were disclosed.

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