CASE REPORT

Atypical Henoch-Schonlein purpura? Consider polyarteritis nodosa!

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Accepted 3 February 2014

SUMMARY

We describe the case of a previously healthy 12-year-old boy admitted to a tertiary paediatric centre with the clinical diagnosis of Henoch-Schonlein purpura (HSP). Symptoms on admission included a generalised rash, colicky abdominal pain, hypertension, proteinuria and fresh rectal bleeding. Abdominal pain and distension worsened and serial ultrasounds suggested bowel ischaemia. He underwent repeat laparotomy and bowel resection, with slow improvement after the second laparotomy. The severity of systemic involvement (gastrointestinal, cardiac, renal and skin) made the initial diagnosis of HSP questionable. Immunohistochemistry of skin biopsies was negative for HSP. Histopathology of the bowel specimen revealed features of necrotising small and medium vessel vasculitis in keeping with polyarteritis nodosa.

BACKGROUND

Henoch-Schonlein purpura (HSP) is the most common systemic vasculitis in children and can involve multiple organs such as skin, kidneys, joints and the gastrointestinal (GI) tract. The disease is usually self-limiting with an excellent prognosis. Abdominal pain in children with HSP is common. It is a manifestation of the vasculitis affecting the GI tract. Less frequently, children with HSP may even present with intussusception of the bowel. Usually however, dramatic complications are rare and major abdominal surgery as described in this case is unusual in HSP. Polyarteritis nodosa (PAN) is a necrotising vasculitis of small-to-medium-sized vessels. Similar to HSP it can manifest itself with a variety of symptoms, including a wide spectrum of GI complications. We report the case of a 12-year-old boy who presented with features suggestive of HSP but also with dramatic systemic organ involvement. He was finally diagnosed with PAN.

We aim to raise awareness of the possible overlap in symptoms of children at initial presentation. A high level of suspicion can prevent a delay in the diagnosis of this severe vasculitis and prompt appropriate treatment.

CASE PRESENTATION

A previously healthy 12-year-old boy was transferred to a tertiary paediatric centre 2 weeks after he had been diagnosed with HSP in a district general hospital. The initial diagnosis had been made clinically on the presence of a generalised maculopapular erythematous rash and colicky abdominal pain. The boy had no medical or family history of significance and an unremarkable social history.

On admission to the tertiary unit he was tachycardic and hypertensive with severe colicky abdominal pain, proteinuria and fresh rectal bleeding. The generalised maculopapular rash continued to be present; his abdomen was soft and mildly tender. Full blood count and biochemical profile were unremarkable; and despite the proteinuria, urea and creatinine were within normal limits (haemoglobin 136 g/L; white cell count $10.9 \times 10^9/L$; urea 4.6 mmol/L; creatinine 58 $\mu\text{mol/L}$). C reactive protein (CRP) was mildly raised at 19 mg/L.

The working diagnosis of HSP was adopted; the history taken together with the rash and abdominal pain seemed typical. The paediatric team instigated medical treatment with oral prednisolone (60 mg three times a day) and antihypertensives, but the boy's clinical condition deteriorated over the next few days. He developed marked abdominal distension and worsening abdominal pain. In addition, he became seriously hypoxic. Blood tests now demonstrated a marked leucocytosis ($40.4 \times 10^9/L$) with left shift and a CRP of 113 mg/L. Proteinuria and haematuria persisted and he developed hypoalbuminaemia. Of note, urea and creatinine remained normal throughout.

As serial ultrasounds showed progressive changes in the small bowel, bowel ischaemia was suspected and the boy proceeded to theatre for exploratory laparotomy. Fifty-two centimetres of small bowel with extensive patchy necrosis were resected. The remaining small bowel showed small areas of ischaemic changes but demonstrated active peristalsis and good blood supply. There was no perforation and no free pus. The colon and other abdominal organs were unremarkable. A large unilateral pleural effusion which had been an incidental finding on the abdominal ultrasound scans was managed with chest drain insertion at the same time. Postoperatively he was started on total parenteral nutrition.

Despite surgical intervention clinical improvement was not sustained. The spasmodic abdominal pain recurred and worsened. It was not controlled with high doses of regular morphine and fentanyl patches and he did not tolerate oral diet. During this time he developed a second pleural effusion on the contralateral side which required a further chest drain insertion. His constantly elevated blood pressure was only partially controlled with a combination of different antihypertensives. The steroid therapy was changed to soluble prednisolone, without any effect on his symptoms. He developed localised areas of subcutaneous oedema and a new purpuric, non-papular rash (figure 1).

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To cite: Braungart S, Campbell A, Besarovic S. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201764

Figure 1 Skin rash on feet and ankles.

The unusual severity and generalised organ involvement led to a review by the paediatric rheumatologist. As a consequence, the steroid therapy was changed to 25 mg intravenous hydrocortisone three times daily, and blood tests for autoantibodies requested. However, all of these ($\beta 2$ glycoprotein, antimitochondrial antibodies, antinuclear antibodies, antismooth muscle antibodies, antiliver-kidney microsomal antibodies, gastric parietal cell antibodies, antinuclear antibodies, perinuclear antineutrophil cytoplasmic antibodies (ANCA), cANCA) were negative/normal. At this point the pathologists advised against obtaining further biopsies; they were certain the diagnosis could be made from the bowel specimen. The intravenous steroid therapy was increased even further to 50 mg hydrocortisone in the morning and 25 mg both at lunch time and in the evening without effect.

Triggered by the non-resolving abdominal pain a CT scan was performed which did not identify any discrete problem, but clinical suspicion was high for on-going bowel ischaemia. Hence, the boy proceeded to a second laparotomy. This time a significant stricture in the small bowel at a marked distance from the previous anastomosis was resected together with further 60 cm of ischaemic small bowel (figure 2A,B). Primary anastomosis was performed. The previous anastomosis and rest of small and large bowel were found to be intact and viable.

INVESTIGATIONS AND TREATMENT

Following the second laparotomy, the boy was restarted on a light diet and started mobilising. This time, there was no recurrence of the abdominal pain, and he continuously improved clinically. Steroid therapy was decreased to 30 mg prednisolone once daily. Although his abdomen stabilised, the rash and subcutaneous oedema still continued to flare up. He developed a grossly swollen left upper arm but no thrombosis was demonstrated on Doppler ultrasonography. His total parenteral nutrition was weaned slowly. He was discharged home on oral prednisolone (30 mg once daily) and Atenolol 8 weeks after onset of his symptoms. Hypertension, haematuria and proteinuria persisted. No involvement of the kidneys could ever be demonstrated on renal ultrasound, renal artery Dopplers were normal. An echocardiogram was unremarkable, with no coronary artery aneurysms present. The spleen and liver remained unaffected. The hypertension was finally attributed to the systemic vasculitis.

The histopathological diagnosis obtained from the bowel specimen at the regional tertiary centre was not clear and the specimen was sent to London for further opinion. It was decided

Figure 2 (A) Severely oedematous, partially ischaemic bowel with patchy necrosis. (B) Small bowel stricture.

that skin biopsies should be obtained in the meantime. These were negative for IgA on direct immunofluorescence which made the diagnosis of HSP even more unlikely.

The final histopathology report of the bowel specimen by the national centre in London diagnosed small and medium necrotising arteritis in keeping with PAN (figure 3).

OUTCOME AND FOLLOW-UP

The boy is now being treated for PAN at the regional rheumatology centre, where he is currently undergoing pulsed intravenous cyclophosphamide and methylprednisolone therapy until remission is evident. His hypertension is managed with a combination of amlodipine, atenolol and aspirin, and he remains on 30 mg prednisolone once daily.

Figure 3 Histopathology of the small bowel revealing necrotising small vessel vasculitis.

Table 1 Criteria for diagnosis of childhood PAN published 2010 by the European League Against Rheumatism/Paediatric Rheumatology International Trials Organization/Paediatric Rheumatology European Society^{7 9}

Criterion	Feature
Mandatory either of the following two	
Histopathology	Necrotising vasculitis evident in small-sized or medium-sized arteries
Angiographic abnormalities	Angiography: aneurysm, stenosis or occlusion of a small-sized or medium-sized artery (not due to fibromuscular dysplasia or other non-inflammatory causes; conventional angiography preferable)
Plus at least one of the following	
Skin involvement	Livedo reticularis, tender subcutaneous nodules, deep/superficial skin infarctions
Muscle involvement	Myalgia or muscle tenderness
Hypertension	Systolic or diastolic blood pressure >95% for height
Peripheral nerve involvement	Sensory peripheral neuropathy
Renal involvement	Proteinuria >0.3 g/24 h or >30 mmol/mg of urine albumin/creatinine ratio on a spot morning sample; haematuria or red blood cell casts >5 per high-power field or red blood cell casts in the urinary sediment or ≥2 on dipstick

PAN, polyarteritis nodosa.

DISCUSSION

HSP is the most common vasculitis in children, with an incidence of about 1:10 000, but unknown aetiology. It presents as a systemic vasculitis, classically with purpura of the lower extremities, joint pain or swelling, GI and renal symptoms. The disease is usually self-limiting with an excellent prognosis.^{1 2} However, other types of vasculitis which are much more severe—such as PAN—can present in a similar way.

Abdominal pain is a common complaint of children with HSP and frequently leads to surgical review.³ The pain is usually periumbilical, colicky in nature and non-specific. In most cases it responds to simple analgesia. The most common abdominal complication requiring surgical intervention in HSP is intussusception—it occurs in about 5% of patients.⁴ Unlike in idiopathic intussusception, the location is most often ileoileal, and more frequent in children >3 years of age. Other severe abdominal complications of HSP include intestinal ischaemia and perforation, and GI bleeding.⁵ However, these are very rare. The generalised severity of symptoms described in the case presentation above was not typical of HSP.⁶ Although the abdominal manifestations were potentially attributable to HSP, the cardiac involvement (hypertension), large recurring pleural effusions and non-resolving symptoms over months were really unusual which is why the initial working diagnosis had to be revised.

PAN is a vasculitis with predominantly small-to-medium-sized blood vessel involvement. It is much rarer in children than HSP but can manifest itself with multiorgan involvement affecting basically any organ.⁷ The organs most often affected are skin, renal and GI tract. Cardiac involvement can manifest itself with hypertension, or even ischaemic heart disease.⁴ If not diagnosed and treated early, PAN has a much higher morbidity. Peak age of onset in children is around 9 years.⁸ Criteria for diagnosing childhood PAN were published recently by the European League Against Rheumatism/Paediatric Rheumatology International Trials Organisation/Paediatric Rheumatology European Society (see table 1).^{5 9}

Children with PAN and severe GI involvement such as acute abdomen have been reported to have a high mortality rate. A case series reported by Venuta *et al*¹⁰ reported eight patients with PAN and acute abdomen, and a mortality rate of 50%. A recent case report by Gomes *et al*¹¹ described the case of two boys with severe cramping abdominal pain, and findings of extensive intestinal necrosis as initial presentation of PAN. These children died following laparotomy despite treatment with multiple immunosuppressants. Unlike in HSP where treatment with steroids remains controversial, corticosteroids are of paramount importance in the treatment of PAN.^{4 12 13} Doses of

at least 1–2 mg/kg/day with or without pulsed corticosteroids are recommended; sometimes cyclophosphamide is needed in addition to that.⁴

The case we report above is unusual, not only because of the final diagnosis of PAN which is uncommon in this age group, but because the boy's initial presentation was suggestive of HSP. However, the unusual severity and generalisation of organ involvement—especially organs not normally affected by HSP—made the initial diagnosis questionable. The final diagnosis of PAN from the bowel specimen was difficult to confirm, but finally obtained by a specialist centre which allowed the instigation of appropriate treatment.

With this report we would like to raise awareness of the possibility of early PAN mimicking HSP. A high level of suspicion is required to distinguish the disease from HSP at an early stage. However, in cases of severe systemic organ involvement, it is extremely important to consider this potentially fatal systemic vasculitis as a differential diagnosis. Multidisciplinary management is paramount to instigate appropriate investigations and treatment in a timely fashion.

Learning points

- ▶ Henoch-Schönlein purpura (HSP) is the most common form of vasculitis in childhood and can present with a variety of symptoms.
- ▶ Extraordinary severity and generalisation of symptoms—especially if non-resolving and involving organs not usually affected by HSP—should raise the suspicion of other types of vasculitis.
- ▶ Polyarteritis nodosa is a necrotising vasculitis of small-sized and medium-sized vessels that can mimic the clinical presentation of HSP.
- ▶ A high level of suspicion is important for correct diagnosis.
- ▶ Multidisciplinary management is crucial to ensure the best possible outcome for children affected.

Contributors SBra conceived the case report, performed the literature search and wrote the article. AC and SBes critically reviewed the article and helped with discussion of the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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