

CASE REPORT

Hypertension in the absence of urinary abnormalities - An unusual presentation of anaphylactoid purpura

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SUMMARY

Henoch-Schonlein Purpura (HSP) or anaphylactoid purpura, currently named IgA vasculitis is the most common form of systemic vasculitis in children. In adults and young infants, HSP tends to have atypical presentations with higher rates of severe gastrointestinal problems and delayed renal complications. While hypertension is a known complication of HSP nephritis, it is rarely seen in individuals with normal renal function and urinary findings. We report a case of a 7-year-old boy with HSP, who presented with abdominal pain and severe hypertension without other features of glomerulonephritis.

CASE PRESENTATION

A 7-year-old well and thriving boy was referred for intermittent, colicky abdominal pain for 2 weeks. He complained of periodic headache and was noted to be persistently hypertensive during his first clinic visit. There were no preceding respiratory symptoms or fever. His urine output was good and there was no frank haematuria. He denied any joint pain or swelling, and there was no rash. There was no known history of trauma, recent travel or ill contact. His immunisations were current, and he received no medications prior to admission. His father was diagnosed with hypertension at the age of 30, currently on treatment and follow-up.

Physical examination revealed he was afebrile with a heart rate 110 beats per minute and blood pressure 155/95mmHg. His peripheries were warm with good peripheral pulses and brisk capillary refilling time. Hydration status was good, and his oropharynx was clear. His pupils were equal, reactive and fundoscopic examination was normal. He had generalised abdominal tenderness but no ascites. There was no limb oedema, joint swelling or rash. Systemic examination was otherwise unremarkable.

A full evaluation was undertaken to look for the cause of hypertension and acute abdomen with an early referral initiated to the surgical team. Blood investigations showed haemoglobin 14.3g/dL, white blood cell 20.6×10^9 , platelet 794×10^9 with elevated ESR at 54mm/h. Serum electrolytes, urea and creatinine were normal. He had normal serum amylase and liver enzymes with albumin 35g/L. ASOT, ANA were negative and there was no hypocomplementemia. Cultures for patient's blood, urine and throat swab were negative. Serial urinalysis, urinary

vanillylmandelic acid, total catecholamines and 17-ketosteroids were normal but plasma renin level was elevated 83.7mIU/L recumbent (normal range: 4.2-59.7). Echocardiography did not show long-standing hypertensive effects or coarctation of aorta. Renal ultrasound and doppler revealed normal size kidneys, good blood flow through the vessels with no structural anomalies. Adrenal glands were not enlarged. Abdominal scan showed minimal free fluid.

His abdominal pain and hypertension persisted despite hydration. Antihypertensive therapy was instituted with oral nifedipine, but his blood pressure remained intermittently elevated at 140-158/88-120 mmHg during the first 3 days of hospitalisation. He required regular doses of nifedipine 0.5mg/kg every 4-6 hour to control his hypertension. One week later in the ward, he developed macular papular rash over both ankles and ears. He was ambulating well and did not report having any joint pain or swelling. Within days, the rashes evolved to prominent, palpable purpuric papules and he passed haeme-positive stools. A repeated abdominal ultrasound reported worsening diffuse thickened loops of small bowel with sluggish movement and increasing pelvic fluid. Skin biopsy showed dense perivascular neutrophilic infiltration admixed with nuclear dusting, endothelial swelling and prominent red blood cells extravasation. Immunofluorescence study demonstrated IgA deposition in the vessels wall and a positive serum cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA) was detected by direct immunofluorescence. The diagnosis of HSP was made and oral prednisolone (1mg/kg/day) was initiated. Repeated serum chemistries and blood counts were likewise unremarkable along with the serial monitoring of urinalysis samples. His blood pressure improved immediately following the commencement of steroid therapy and his abdominal pain gradually resolved. Over the subsequent 10days, his blood pressure was well controlled at 95-110/60-75mmHg and antihypertensive medication was discontinued for 48 hours prior to his discharge. He was given tapering doses of prednisolone over 4 weeks. Several evaluations to 6 months after his admission also revealed blood pressures 99-102/62-68mmHg, with normal urinalyses and renal function.

DISCUSSION

The diagnostic criteria for HSP include palpable purpura on lower limbs as a mandatory criteria and the presence of at least one of the following: diffuse abdominal pain, arthritis or arthralgia,



Fig. 1: Maculo-papular rash on both feet and ankles.

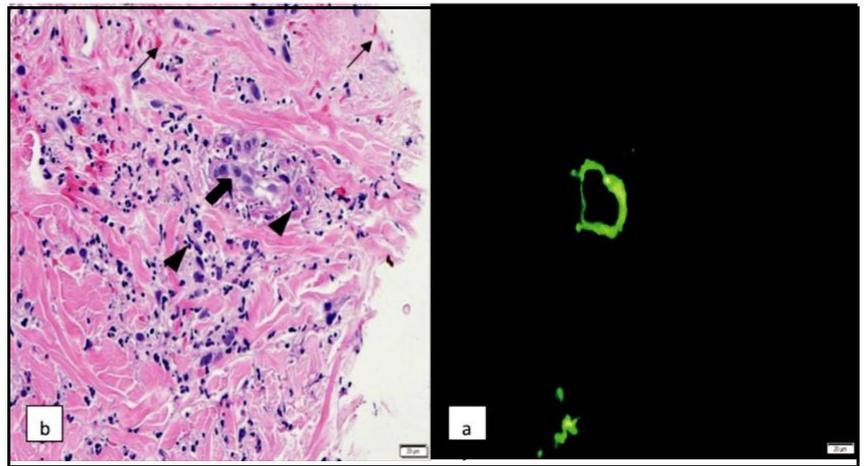


Fig. 2: a) Histopathology of the cutaneous lesion revealing endothelial cell swelling (thick arrow) with neutrophilic infiltrate in the blood vessel wall (arrow head) and red blood cell extravasation (thin arrow). b) Immune deposition of IgA at the blood vessel.

haematuria or proteinuria and vascular deposits of IgA. Atypical features of abdominal pain and hypertension in the absence of cutaneous and joint manifestations at the initial phase may impose diagnostic issues among clinicians. Moreover, the presence of severe hypertension without evidence of renal or urinary abnormalities is uncommon.

Our patient presented with an aberrant sequence of initial abdominal pain and severe neurological symptoms including headache and hypertension. The appearance of purpuric rash was also delayed until the end of his clinical course as opposed to the typical presentation at the onset of illness. He also developed other complications including intestinal vasculitis and hypertension without evidence of renal disease. In the absence of characteristic rash and overt arthritis, persistent hypertension and abdominal pain during the early phase of HSP is rare. This resulted in an initial diagnostic dilemma and delayed steroid initiation.

Hypertension is a known complication associated with renal involvement in HSP and its presence is almost universally allied with other evidence of nephritis. Renal involvement usually occurs within 3 months of the onset of the rash and in children, it is usually found within the first month of illness. Common presentations include haematuria (gross or microscopic), proteinuria, nephrotic syndrome, mixed nephrotic-nephritic picture or rapidly progressive glomerulonephritis. Isolated hypertension in HSP without renal involvement is rare with very few cases reported. Among the cases described previously, a patient had severe abdominal pain and protein-losing enteritis accompanying hypertension.¹ The authors postulated the presence of hypoproteinemia as a possible trigger to renin hyper-production, a factor which was not present in our patient. Among other reports in

the literature was an illustrated case of HSP with no evidence of renal involvement, who presented with significant hypertension which was resistant to treatment with β -adrenergic blocking agents and vasodilators. Initiation of captopril and corticosteroid therapy resulted in prompt resolution of hypertension suggesting a renin-angiotensin-mediated mechanism for hypertension. The hypertension recurred with tapering doses of steroids, only to respond to higher doses of glucocorticoids.²

Our patient had persistent hypertension with normal urinalyses and serum renal indices throughout his course of illness. In fact, his blood pressure remained elevated despite the initiation of nifedipine and on occasions when he did not experience any abdominal pain. His blood pressure only showed significant improvement upon initiation of prednisolone. While established glomerular disease in HSP may respond poorly to glucocorticoid therapy,³ acute reno-vascular hypertension in this small-vessel vasculitis syndrome may be responsive to corticosteroids in at least a subset of children with this disorder. Proof of reno-vascular cause in this patient can be deduced from the presence of tachycardia and persistent hypertension, absence of glomerulonephritis, elevated plasma renin levels and response of patients to glucocorticoid treatment. This also suggests that renal arteriolar disease may occur in the absence of any evidence of glomerulonephritis in HSP. Biopsy confirmation of normal renal histology would be of value, but this was not indicated on clinical grounds as this patient had no renal or urinary abnormalities. Among the other non-renal aetiologies for hypertension is the possibility of excessive adrenal discharge, perhaps from adrenal vasculitis. However, this is only vaguely supported by the presence of tachycardia and hypertension in this patient. His urine examination for steroids and catecholamines were normal. Last but not least, positive markers for ANCA are rarely found in HSP as opposed to pauci-immune small vessel vasculitis. Albeit there have been cases reported in the past of HSP with ANCA positivity.⁴

CONCLUSION

We propose that the present case demonstrates an atypical presentation of anaphylactoid purpura or HSP. Our case also highlights that hypertension can occur in an occasional patient with HSP without evidence of renal disease, either decreased renal function or urinary abnormalities. Thus, those providing immediate care should be reminded of the potential multisystemic involvement of HSP in the paediatric patient population.

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DECLARATION OF CONFLICTING INTERESTS

The author(s) declared no potential conflicts of interest with respect to the authorship and/or publication of this article.

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