

Severe Mitral Regurgitation in a Child With Henoch-Schönlein Purpura and Pulmonary Hemorrhage

Global Pediatric Health
Volume 4: 1–4
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DOI: 10.1177/2333794X17726940
journals.sagepub.com/home/gph



Cornelius A. James, MD^{1,2}, Ismael Gonzalez, MD^{1,2}, Paras Khandhar, MD^{1,2}, and Bishara J. Freij, MD^{1,2}

Abstract

Introduction: Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood. The classic triad of HSP consists of nonthrombocytopenic purpura, arthritis/arthralgia, and gastrointestinal complaints. Pulmonary hemorrhage and cardiac involvement are rare complications of HSP. **Case Report:** We report the case of a 10-year-old girl with HSP complicated by both severe mitral regurgitation and pulmonary hemorrhage. **Discussion:** HSP is typically a self-limited illness with an excellent prognosis in children. Pulmonary hemorrhage is a rare complication that increases morbidity and mortality; it generally indicates the presence of severe vasculitis. Cardiac involvement in HSP is extremely rare and associated with a poor prognosis. **Conclusion:** Cardiac involvement in HSP may be more common than believed. Because of the increased morbidity and mortality associated with HSP complicated by pulmonary hemorrhage and cardiac involvement, it is important for clinicians to be aware of these potential complications.

Keywords

Henoch-Schönlein purpura, pulmonary hemorrhage, carditis, leukocytoclastic, mitral regurgitation

Received May 5, 2017. Accepted for publication July 17, 2017.

Introduction

Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood. HSP is a leukocytoclastic, small vessel vasculitis (SVV) characterized by immunoglobulin A (IgA) deposition into the small vessels of multiple organ systems. The classic triad of HSP consists of nonthrombocytopenic purpura, arthritis/arthralgia, and gastrointestinal complaints. Pulmonary hemorrhage and cardiac involvement are rare complications of HSP. We report a case of a 10-year-old child with HSP complicated by both severe mitral regurgitation and pulmonary hemorrhage.

Case Report

A 10-year-old Caucasian girl presented with complaints of progressively worsening rash, fever, and shortness of breath. Evaluation during the preceding 10 days included a normal complete blood count, electrolyte panel, liver function tests, urinalysis, and erythrocyte sedimentation rate (ESR). C-reactive protein (CRP) was 1.2 mg/dL

(normal <0.8 mg/dL), serum IgA 310 mg/dL (normal 52-256 mg/dL), and anti-streptolysin O (ASO) titer 376 IU/mL (normal 0-249 IU/mL). Her primary care physician noted a rash on her posterior thighs, buttocks, and elbows, and she complained of pain in her elbows and knees. HSP was suspected. A temperature of 38.4°C, shortness of breath, dry cough, and poor appetite prompted a visit to the emergency center.

On presentation, her oxygen saturation was 91% on room air, respiratory rate 33/min, and heart rate 133/min. She had dry mucous membranes, pulmonary rhonchi, and a purpuric rash on her upper and lower extremities, buttocks, and back. Her hemoglobin had decreased from 14.6 mg/dL 10 days prior to 10.7 mg/dL. White

¹Oakland University William Beaumont School of Medicine, Rochester, MI, USA

²Beaumont Children's Hospital, Royal Oak, MI, USA

Corresponding Author:

Paras Khandhar, Department of Pediatrics, Beaumont Children's Hospital, 3601 W 13 Mile Rd, Royal Oak, MI 48073, USA.

Email: paras.khandhar@beaumont.org



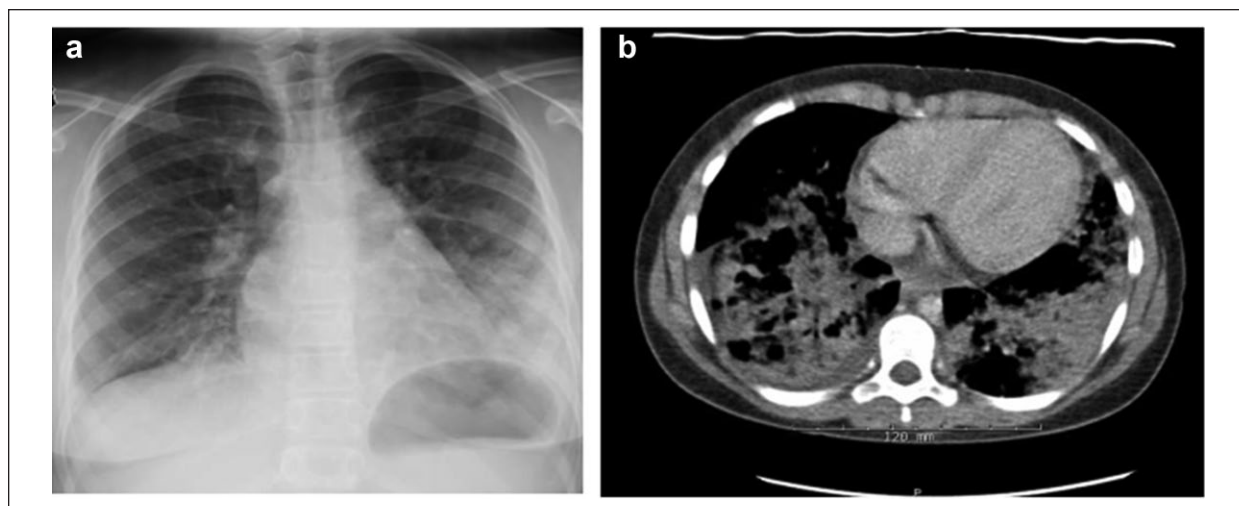


Figure 1. (a) Chest radiograph showing patchy bibasilar opacities. (b) Computed tomography of chest demonstrating bibasilar airspace disease.

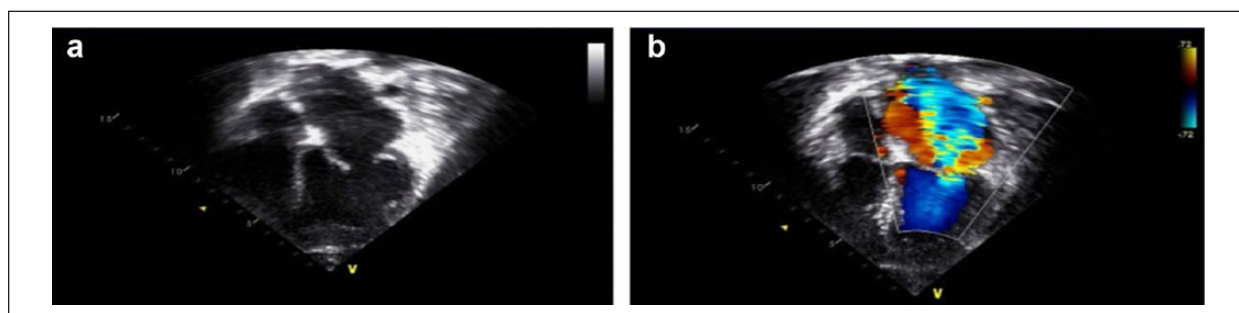


Figure 2. (a) Echocardiogram demonstrating dilated left atrium and left ventricle, and a structurally normal mitral valve. (b) Echocardiogram showing severe mitral regurgitation.

blood cell count was 12.0 bil/L and platelet count 350 bil/L. A chest radiograph showed patchy bibasilar opacities (Figure 1a). Intravenous ampicillin and azithromycin were started.

Shortly after admission she began grunting, had respiratory rates of 38 to 58/min, and a capillary blood gas revealed a pH of 7.50, and $p\text{CO}_2$ 28 mm Hg while on supplemental oxygen. A repeat chest image showed worsening of the bibasilar opacities and a prominent cardiac silhouette. High flow oxygen via nasal cannula was started and antibiotics changed to vancomycin, ceftriaxone, and azithromycin. The ASO titer had increased to 654 IU/mL and an anti-DNase B antibody titer was 187 U/mL (normal 0-170 U/mL). Chest computed tomography demonstrated bibasilar airspace disease, possibly an infectious process, or hemorrhage secondary to vasculitis (Figure 1b). Intravenous methylprednisolone (30 mg/kg/day) was started for treatment of presumed severe SVV.

She developed an apical III/VI holosystolic murmur that radiated to the axilla. Echocardiogram demonstrated a mildly enlarged left ventricular cavity, a normal mitral valve but moderate to severe mitral regurgitation (Figure 2). Her initial b-type natriuretic peptide was 621 pg/mL (normal 0-100 pg/mL). Intravenous furosemide was given to prevent volume overload. The following day an echocardiogram revealed severe mitral regurgitation, a marked increase in the size of the left atrium, and diastolic dysfunction. Milrinone and intravenous epinephrine infusion were started for afterload reduction and worsening hypotension, respectively. Her respiratory status continued to deteriorate prompting endotracheal intubation. During nontraumatic intubation a copious amount of fresh blood was encountered. Intravenous gammaglobulin (1 g/kg/day) was initiated for treatment of severe vasculitis. Repeat CRP was 11.3 mg/dL and ESR 25 mm/h (normal 0-18 mm/h). Throat, tracheal aspirate, and blood cultures were negative.

She gradually improved, and was extubated after 5 days of mechanical ventilation. Milrinone and epinephrine infusions were weaned and an oral prednisone taper was started. Enalapril was initiated for continued afterload reduction. She remained stable on room air and was discharged after completing 10 days of intravenous ceftriaxone and 7 days of intravenous azithromycin. Prior to discharge an echocardiogram showed a moderately dilated left atrium, moderate mitral regurgitation, and mild mitral valve prolapse. At discharge, she continued enalapril and a 4-week steroid taper.

One year after discharge she was asymptomatic and doing well and an echocardiogram revealed mild to moderate mitral regurgitation. The left ventricle and left atrium were normal.

Discussion

HSP is typically a self-limited illness with an excellent prognosis in children. The illness tends to be more severe in adults and is more likely to result in long-term complications. Pulmonary hemorrhage is a rare complication of HSP and increases morbidity and mortality; it generally indicates the presence of severe vasculitis. Dyspnea was the most common manifestation of pulmonary hemorrhage in 17 patients reported by Chen et al.¹ Symptoms may also include chest pain, fatigue, altered activity tolerance, cyanosis, cough, hemoptysis, and anemia. Bilateral fluffy, diffuse, or patchy opacities are the most common imaging findings.¹ Similar to our experience, others have aspirated large amounts of blood from the endotracheal tube during intubation in patients with pulmonary hemorrhage.^{2,3} Our patient had significant respiratory distress, anemia, bilateral patchy opacities on imaging, and a copious amount of blood was encountered during intubation.

Cardiac involvement in HSP is extremely rare and associated with a poor prognosis. There have been reports of carditis associated with HSP. Several investigators consider acute rheumatic fever (ARF) to be the cause of the carditis as many of these patients had both evidence of previous or current group A beta hemolytic streptococcal (GABHS) infection⁴⁻⁶ as well as the typical clinical manifestations (the Jones' criteria) required for the diagnosis.⁷ However, carditis, polyarthritis, arthralgia, fever, and elevated acute phase reactants are all non-specific and occur in other illnesses, including HSP.

HSP in the setting of findings consistent with rheumatic carditis has led some authors to theorize that HSP and rheumatic fever share a common etiologic agent in GABHS. However, these 2 diseases represent 2 very different immunologic or pathogenic processes. HSP is a disease mediated by IgA immune complexes, while

rheumatic carditis is believed to be mediated by molecular mimicry between streptococcal antigens and human proteins leading to autoimmune reactions.

HSP with cardiac involvement occurs more often in adults. Arrhythmia, cardiac necrosis, bundle branch block, and myocardial infarction have been reported in adults with HSP.^{8,9} Lutz et al described 12 cases of HSP with cardiac involvement.⁹ One third of the patients had evidence of streptococcal infection and 5 died, confirming the unfavorable prognosis when HSP is complicated by cardiac involvement. Hung et al described a case of a 2-month-old infant with severe leukocytoclastic vasculitis with endomyocarditis, mitral regurgitation, and pulmonary hemorrhage.¹⁰ Evidence of GABHS infection was not reported in this patient. The authors concluded that cardiac involvement presenting as severe mitral regurgitation is possible in leukocytoclastic vasculitis in children.

Our patient's clinical presentation, including non-thrombocytopenic purpura in dependent areas, arthritis and arthralgia in the setting of elevated serum IgA contributed to a diagnosis of HSP. The presence of pulmonary hemorrhage made it likely that she had severe SVV secondary to HSP. While she did have elevated ASO, CRP, ESR, and anti-DNase B levels, it is more likely that the carditis was secondary to severe small vessel vasculitis and not ARF given the structurally and functionally normal mitral valve on echocardiogram. It is unlikely that our patient had rheumatic carditis, despite satisfaction of the Jones criteria with 1 major (carditis) and 2 minor (elevated acute phase reactants, fever) criteria, and evidence of antecedent GABHS infection.

Conclusion

SVV can cause myocardial and valvular dysfunction that can mimic ARF. Cardiac involvement in HSP may be more common than believed. Because of the increased morbidity and mortality associated with HSP complicated by pulmonary hemorrhage and cardiac involvement, it is important for clinicians to be aware of these potential complications. Distinguishing between ARF and carditis secondary to HSP may have a significant effect on treatment, both acutely and long term, as a diagnosis of acute rheumatic fever commits a patient to long-term prophylactic antibiotics. Caution must be exercised when distinguishing between carditis secondary to severe small vessel vasculitis and ARF in patients with HSP.

Acknowledgments

Donald Gibson, MD, Department of Diagnostic Imaging, Beaumont Children's Hospital, Royal Oak, MI; Beth Kring, BSN, CMN, CCRC, Beaumont Health, Royal Oak, MI.

Author Contributions

CAJ: Contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

IG: Contributed to conception and design; contributed to analysis; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

PK: Contributed to conception and design; contributed to acquisition, analysis, and interpretation; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

BJF: Contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

Declaration of Conflicting Interests

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

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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