

Lumbosacral Edema as an Unusual Presentation of Henoch-Schönlein purpura: A Case Report and Literature Review

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Abstract

Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood. Subcutaneous edema in the extremities is a common finding of HSP, whereas lumbosacral edema is a rare presentation. We report a case of HSP in a six year old boy who visited the emergency department for acute-onset back pain with gait problems. An extensive radiologic workup was performed, but the patient was finally diagnosed with HSP based on knee and ankle edema with a few purpura. We described the clinical, imaging findings, and treatment of the current case and review the literature regarding this rare presentation of HSP.

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Introduction

Henoch-Schönlein purpura (HSP) is the most common vasculitis in children. Clinical tetrad of HSP comprised palpable purpura, arthralgias, abdominal pain and renal disease. Subcutaneous edema (Swelling) is another common manifestation of HSP. The majority of subcutaneous edema was located on the extremities, including hands, ankles and feet.¹ Edema occurring in other locations such as facial, scalp and scrotum is uncommon but well-described.² Subcutaneous edema of the lumbosacral area has been recently reported as an infrequent presentation of HSP.³⁻⁷ This unusual presentation of HSP needs to be well aware because it manifests as acute and severe backache, a symptom that could puzzle physicians. Here, we report a case of HSP with lumbosacral edema that led to concern about the serious condition of the spinal cord. We also review the literature regarding this rare presentation of HSP.

Case Report

A previously healthy six year old boy visited the emergency department (ED) with sudden back pain and refusal to walk. One week before visiting the ED, he was treated for bacterial pharyngitis. He had pain on both calves and knees, but he could walk. At presentation, the patient experienced severe lower back pain and could not stand by himself. There was no trauma. There was tenderness of the knees, calves and back. Soft tissue swelling in the lumbar region with loss of normal lumbar curve was seen on lateral X-ray view of the lumbar spine (Fig 1). There were no overlying skin changes but had a restricted range of motion with forwarding bending. Rest of the neurological findings were normal. MRI of the lumbosacral spine revealed subcutaneous and fascial edema in the lumbar area only (Fig 2).

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There were several purpuras and left knee swelling with bruising on the popliteal region (Fig. 3). All relevant laboratory studies were within normal range. He was ultimately diagnosed with HSP and managed with an oral corticosteroid (2 mg / kg). He started to improve significantly the next day and was asymptomatic after a few days. During the 12-week follow-up period, the patient showed no evidence of HSP-related complications or recurrence.

Fig. 1. A lateral X-ray view of the lumbar spine showed subcutaneous edema and soft tissue swelling (arrow), leading to the loss of the normal lumbar curve.

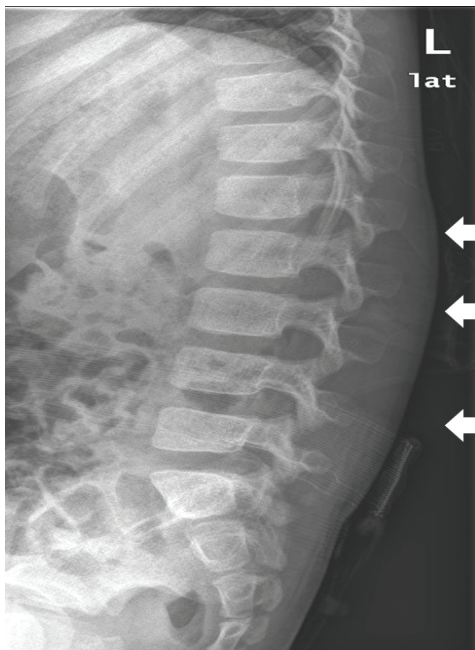


Fig. 2. Magnetic resonance imaging (MRI) of the lumbar spine. Sagittal T2-weighted, fat-suppressed MRI was showing a hyperintense signal in the posterior lumbar region (A). Transverse T1-weighted MRI shows a hyperintense signal on the subfacial region (*), compressing the paraspinal muscles (B).

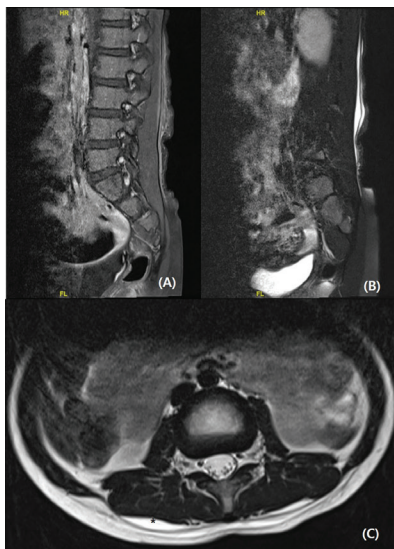


Fig. 3. Swelling with bruises on the popliteal area and purpura (arrow) on the ankle of the left leg.



Discussion

Gait problems, particularly refusal to walk, are commonly encountered in children with HSP if arthralgia / arthritis are accompanied in ankles and knees.⁹ However, HSP may not be the primary disease to exclude when gait problems are accompanied by acute back pain. In our case, acute-onset back pain and refusal to walk made us do the MRI. We could suspect HSP with purpura and knee swelling with arthralgia. These features met with the diagnostic criteria proposed by the European League Against Rheumatism (EULAR) / Paediatric Rheumatology European Society (PReS) / Paediatric Rheumatology International Trials Organisation (PRINTO).^{7,8} Our case shows that HSP should be considered a broad differential diagnosis in patients with atraumatic lower back pain, swelling, and refusal to walk. This case also highlights the importance of careful physical examination for HSP diagnosis. To our knowledge, only five cases of HSP with lumbar edema have been reported (Table 1).^{3,7} Consistent symptoms associated with lumbosacral edema were acute - onset severe back pain and refusal to walk. Back pain was the initial symptom of HSP reported on first to second day of illness, except for one patient.^{3,7} Interestingly, most of these patients whose back pain was an initial symptom were admitted to the hospital via ED,^{3,7} probably because the back pain was sudden and severe. Our patient also had arthralgia in ankles and knees the day before, but he visited the ED for subsequent sudden back pain. Lumbosacral edema with back pain is very unusual as the initial symptom of HSP. However, the diagnosis seemed relatively straightforward in most cases because of concomitant purpura and / or arthralgia.^{3,6,7} Meanwhile, it could be challenging to diagnose HSP when purpura is accompanied by lumbosacral edema alone without other joint symptoms or abdominal pain. Shimizu et al. mentioned that they hesitated to diagnose HSP in a seven year old girl with lumbosacral edema with acute-onset severe left low back with severe tenderness.⁵

In almost all reported cases, extensive radiological evaluation such as CT or MRI was performed to exclude subcutaneous hematoma secondary to potential trauma or blood diseases. Our MRI findings are consistent with previous reports and suggest subcutaneous and fascial edema of the paraspinal muscles.^{3,5} Subfacial fluid collection (below deep fascia) compressing the

underlying muscles was observed in our case (Fig 1B). Similar findings were also reported in Shimizu et al., in which unilateral subfascial fluid collection and ipsilateral low back pain were described.⁵ American football-shaped edema in the posterior lumbar region has been reported in HSP,⁷ supporting the possible involvement of the deep fascia. Fluid collection in the narrow space below the deep fascia might lead to severe pain and

limitation of forwarding bending in our patient. After receiving steroids, our patient's knee and back symptoms improved quickly. All reported cases showed that rapid improvement of back pain with or without medications.³⁻⁷ These findings suggest that lumbar swelling shares a similar mechanism as HSP edema, which commonly leads to swelling of the extremities.⁷

Table 1. Summary of the literature review findings

	Present case	Schaefer et al. 2014 ³	Duman et al. 2016 ⁴	Shimizu et al. 2019 ⁵	Yasumura et al. 2020 ⁶	Winkle et al. 2020 ⁷
Age, yr	6	5	4	7	6	6
sex	male	male	Male	Female	Male	Female
Route of admission	ED	ED	ED	Unknown	unknown	ED
The main symptoms that led to admission	Severe back pain and refusal to walk	Severe back pain and refusal to walk	Increasing abdominal pain	Severe left low back pain	Sudden onset of severe back pain	Low back pain and swelling
The first symptoms of HSP	Calf and knee pain	Non-purpuric erythematous rash	Abdominal pain	Back pain	Back pain, bilateral knee joint pain, and right leg edema	Back pain, joint pain of lower extremities
Symptoms/signs of LS edema	Back pain and refusal to walk/ lumbar swelling and restriction of forwarding bending	Back pain and refusal to walk/ lumbar swelling and limitation of forwarding bending	Back pain and refusal to walk/ lumbar swelling	Back pain/ Severe tenderness and swelling in the left low back region	Back pain/ marked swelling of the lumbar back	Low back pain/ American football-shaped edema on the lower back with moderate tenderness
Day of illness presenting LS edema	2 nd day	1 st day of illness	11 th day of illness	1 st day of illness	Unknown	1 st day of illness
When to observe purpura with purpura	At the same time	Before LS edema	Before LS edema	At the same time	after LS edema	unknown
Arthralgia/joint swelling	(+)	(+)	(-)	(-)	(+)	(+)
Abdominal pain	(-)	(+)	(+)	(+)	(+)	(-)
Kidney involvement	(-)	Proteinuria	(-)	unknown	unknown	Hematuria
Evaluation tools for LS edema	X-ray/MRI	MRI	CT/ MRI	MRI	CT/MRI	X-ray
Treatment	steroid	NSAID	Oral steroid	No medication	No medication	NSAID
Response to treatment	Improve following 72 hours	Improve following 24 hours	Improved following 48 hours	Improved following days	Improved	Improved
Follow-up	12 weeks, no symptoms of HSP	12weeks, no symptoms of HSP	A month, a relapse of rash only	unknown	2 months, symptoms	48 hours, symptoms free

Abbreviation: ED, emergency department; LS, lumbosacral; MRI, magnetic resonance imaging; CT, computed tomography; NSAID, non-steroid anti-inflammatory drug; HSP, Henoch-Schönlein purpura

Conclusions

The present case shows that paediatricians need to know lumbosacral edema as a rare HSP presentation that manifests with abrupt-onset back pain and gait problems. Detailed history taking and careful physical exam can provide critical clues for HSP suspicion.

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