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Unusual Lower Back Pain and Swelling Associated with IgA Vasculitis: A Case Report with Review of Literature

Hiro Matsukura[1,*], Shokei Murakami[2]

[1]Department of Pediatrics, Saiseikai Takaoka Hospital, Takaoka, Japan
[2]Department of Pediatrics, University of Toyama, Faculty of Medicine, Toyama, Japan
*Corresponding author: matsukura@5x2.de

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Abstract IgA vasculitis is the most common childhood vasculitis. Subcutaneous edema of the extremities is a common finding in IgA vasculitis, but posterior lumbar subcutaneous edema is a rare complication. A four-year-old boy presented to the emergency department with sudden onset of low back pain, swelling, and refusal to walk. The patient had no history of obesity, spinal trauma, or cardiac, hepatic, or renal dysfunction causing the edema. No prior infection was confirmed. The spine was tender to palpation and swollen over the thoracolumbar vertebrae. Arthralgias of the knees and ankles with a purpuric rash on the lower extremities confirmed the diagnosis of IgA vasculitis. There was no abdominal pain and the urinalysis was normal. Normal plasma factor XIII activity may reflect mild disease or the absence of abdominal pain. The posterior lumbar subcutaneous edema improved immediately with acetaminophen. The leakage of blood vessels into the surrounding tissues due to the vasculitic inflammatory process may result in the posterior lumbar subcutaneous edema. We conducted a literature review of previous case reports of posterior lumbar subcutaneous edema. Most of the patients presented directly to the emergency department and refused to ambulate. None of the patients had superimposed petechiae and purpura over the thoracolumbar spine. All but one patient received corticosteroids or nonsteroidal anti-inflammatory drugs. Lumbar pain and swelling improved completely without recurrence. Posterior lumbar subcutaneous edema in the setting of IgA vasculitis was rare and was a self-limiting symptom of limited duration.

Keywords: *Henoch-Schönlein purpura, IgA vasculitis, plasma factor XIII, posterior lumbar subcutaneous edema*

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1. Introduction

IgA vasculitis (IgAV), formerly Henoch-Schönlein purpura, was the most common vasculitis in children. [1,2,3,4,5,6,8,9] The etiology was unknown, but IgAV typically followed an upper respiratory infection. [1,3,5,8] IgAV was characterized by a tetrad of abdominal pain, nonthrombocytopenic purpura, arthritis, and renal disease. [1,2,3,5,6,7,8,9] Subcutaneous edema, especially in the hands, ankles, and feet, is a common finding in IgAV. [1,3,4,5,6,7,8,9] Edema at other sites is uncommon. Face, scalp, peno-scrotal and labial edema have been reported. [1,3,8,9] Facial edema is most common in children under the age of 2. [3] Herein, we presented an unusual case of posterior lumbar pain and swelling associated with IgAV along with a literature review highlighting the clinical features of posterior lumbar subcutaneous edema (PLSE) associated with IgAV.

2. Case Report

A previously healthy Japanese four-year-old boy presented to the emergency department with sudden onset of severe back pain, swelling, and refusal to ambulate. One week prior to presentation, a rash had developed on the lower extremities. The patient was fully immunized and had an unremarkable past medical and family history. The patient was not obese with normal Kaup index of 15.6 (body weight 16 kg, height 101 cm). The patient had no history of spinal trauma or cardiac, hepatic, or renal dysfunction causing edema. Temperature was 37.5°C, the blood pressure 102/53 mmHg, the pulse 107 beats per minute, the respiratory rate 20 breaths per minute, and 99% pulse oximetry on room air. Multiple, raised, round, pruritic, and painless papules were present on the bilateral lower legs (Figure 1a). Left calf and ankle were swollen and tender. The abdomen was soft without tenderness. Dorsal examination revealed a normal thoracic spine with marked symmetrical soft tissue swelling in the lumbar region obscuring the spinous processes and with loss of normal lumbar curvature (Figure 1b). There was severe tenderness to palpation over the bony processes and paraspinal muscles without overlying skin changes. Neurological examination was normal.

Table 1. Baseline laboratory investigations of the child

Parameters	Results
WBC (10^3/mm^3)	9.1
N/L/M/E (%)	57.9/31.9/5.7/4.1
RBC (10^6/mm^3)	4.35
Hemoglobin (g/dL)	11.8
Hematocrit (%)	33.4
Platelet (10^3/mm^3)	357.0
C-reactive protein (mg/dL)	0.09
Total protein/albumin (g/dL)	6.7/4.4
Total bilirubin (mg/dL)	0.33
AST/ALT (U/L)	22/10
Alkaline phosphatase (U/L)	192
Serum GGT (U/L)	9
LDH (U/L)	208
Urea/creatinine (mg/dL)	15.3/0.31
Sodium/potassium/chloride (mmol/L)	138/3.8/106
Calcium/phosphate (mg/dL)	9.8/4.6
Creatinine kinase (U/L)	102
Prothrombin time (INR)	0.98
Activated thromboplastin time (sec)	30.1
Fibrinogen (mg/dL)	308
Fibrinogen degradation products (µg/mL)	2.5
D-dimer (µg/mL)	1.17
Plasma factor XIII (%)	119
Urinalysis	
Proteinuria	Negative
Glucose	Negative
Occult blood test	Negative
Sediment	WBC 1-4/HPF RBC 0-1/HPF

WBC: white blood cells, N/L/M/E: Neutrophils/ lymphocytes/ monocytes/ eosinophils, RBC: red blood cells, AST: Aspartate aminotransferase, ALT: Alanine aminotransferase, GGT: Gamma-glutamyl transferase LDH: Lactate dehydrogenase

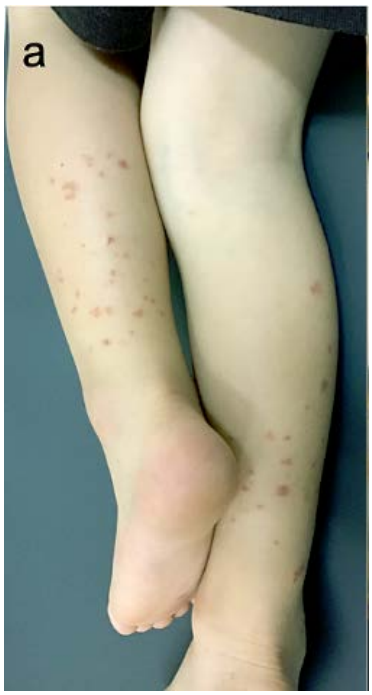

Figure 1a. Raised purpuric rash is present on the dorsal aspect of the lower extremities

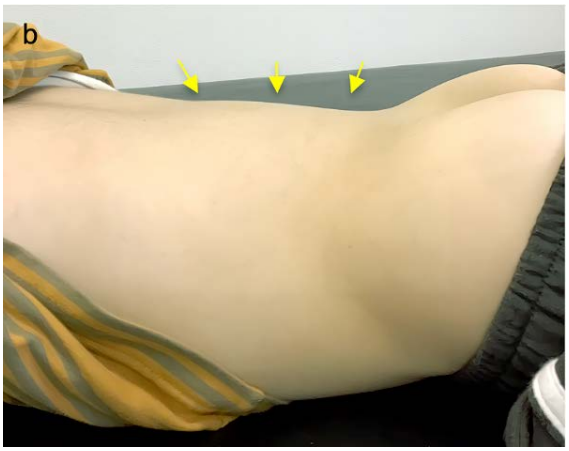

Figure 1b. Photograph of the patient's lower back shows lumbar soft tissue swelling as indicated by the arrows. Upon arrival at the emergency room, the patient was found to have a swelling in his lower back the size of an American football. The photo taken the next day showed that the swelling had decreased but was still present

Laboratory studies Table 1 included a normal complete blood count, C-reactive protein, creatinine phosphokinase, prothrombin time, and partial thromboplastin time. Plasma factor XIII activity was 119% (normal 70-140%). Urinalysis was normal. Details of the clinical laboratory data are presented in Table 1. The patient was diagnosed with IgAV and treated with acetaminophen. The lumbosacral pain and swelling resolved promptly, but the left leg arthralgia persisted for ten days. The patient was seen fourteen days later and had no abdominal pain and no recurrence of back pain and edema. The patient will be followed closely for the development of abdominal pain and renal disease.

3. Discussion

We found two clinically important points. PLSE associated with IgAV was rare and caused sudden severe low back pain.

First, localized subcutaneous edema was common in IgAV, but PLSE was rare. Quincke's edema and hereditary angioedema are characterized by the appearance of localized edema. Quincke's edema often complicated cases of IgAV. [1,3,5,6,7,8] Deepashree et al. reported a seven-year-old girl with unilateral eyelid angioedema associated with IgAV. [10] Angiotensin converting enzyme (ACE) inhibitors cause acute angioedema seen in the emergency department, but the patient was not taking ACE inhibitors. [11] Quincke's edema usually involves the face, lips, and/or tongue and may occur on the dorsum of the hands or feet, genitals, or abdomen, but the lumbar region is rarely involved. [12] Hereditary angioedema (HAE) is a rare disorder. Complement tests including C3, C4, CH50, and C1 inhibitor, which would allow a more accurate differentiation from HAE, were not performed. [11] No other family members of the patient were affected. The patient had pruritus without recurrent episodes of angioedema. HAE was unlikely in this patient.

Young first reported a five-year-old boy with PLSE, but the author focused on the intestinal obstruction and did not describe the clinical course of the PLSE. [13] Table 2

compares our case with ten similar cases previously reported in the literature. [1,2,3,4,5] [6,7,8,9] Of these patients, eight were male and three were female with a median age of five years. IgAV developed classically after the upper respiratory tract infection. [1,3,5,8] Ziyara et al. reported a twelve-year-old boy who developed IgAV with a novel coronavirus disease 2019. [14] The previous infection was not identified in the patient. Triggering events for PLSE were observed in three patients. Patient 1 was triggered by trampoline play, patient 4 by swimming, and patient 5 by influenza vaccination. [2,4,5] The patient had been doing well when suddenly, without any triggering event, severe back pain, swelling and a refusal to walk developed. The seventh day interval between IgAV and PLSE in this patient was slightly longer than in the other cases (median second day). PLSE developed on the same day as the appearance of IgAV in three out of ten patients. Three days of disappearance of PLSE in the patient was similar to those of other cases. The classic tetrad of palpable purpura, abdominal pain, arthralgia, and renal involvement was seen in ten, eight, eight, and three patients, respectively.

Table 2. Review of children with posterior lumbar subcutaneous edema associated with IgA vasculitis

Author, year of publication, place	Age/ gender	Interval* (day)	Duration# (days)	Emergency department	Refusal to walk	Abdominal pain	Arthralgia	Purpura	Urinalysis	Factor XIII (70-140%)	Treatment	Image diagnosis
Robson and Leung, 1995, USA[2]	9 Male	6th	6	No	+	+	+	+	Hematuria Proteinuria	N/A	Steroids	Radiology
Schaefer and Soprano, 2014, USA[1]	5 Male	1st	2	Yes	+	+	-	+	Proteinuria	N/A	NSAIDs	MRI
Duman *et al*., 2016, Turkey[3]	4 Male	11th	3	Yes	+	+	-	+$	Normal	N/A	NSAIDs	CT/MRI
Shimizu *et el*., 2019, Japan[4]	7 Female	1st	3	No	-	+	-	+	N/A	51%	Steroids	MRI
Bouchard and Sidlow, 2020, USA[5]	4 Female	2nd	2	Yes	+	+	+	+	Normal	N/A	NSAIDs	MRI
Winkler *et al*., 2020, USA[6]	6 Female	1st	5	Yes	+	-	+	+	Hematuria	N/A	NSAIDs	Radiology
Yasumura *et al*., 2021, Japan[7]	6 Male	1st	2	No	+	+	+	+	Normal	Decreased	None	CT/MRI
Marcia and Parodi, 2022, Italy[8]	3 Male	3rd	3	Yes	+	+	+	-	Normal	53%	Antihistamines	None
Marcia and Parodi, 2022, Italy[8]	4 Male	10th	5	No	N/A	+	+	+	Normal	32%	Steroids	None
Yeom *et al*., 2022, South Korea[9]	6 Male	2nd	3	Yes	+	-	+	+	Normal	N/A	Steroids	Radiology/ MRI
Our case	4 Male	7th	3	Yes	+	-	+	+	Normal	119%	NSAIDs	None

*Number of days from the onset of IgA vasculitis to the appearance of posterior lumbar subcutaneous edema, #How many days after posterior lumbar subcutaneous edema disappe
$Purpura developing one day after admission, N/A: not available, NSAIDs: nonsteroidal anti-inflammatory drugs, MRI: magnetic resonance image, CT: computed tomography

Normal platelet counts and coagulation studies differentiated IgAV from other conditions associated with petechiae and purpuric rashes including septicemia, immune thrombocytopenia purpura, hemolytic uremic syndrome, leukemia, and coagulopathies in our patient. Skin biopsy was not performed. The diagnosis did not require all four elements. Our patient fulfilled at least two of the EULAR/PRINTO/PRES criteria (palpable purpura and arthritis or arthralgia), which was compatible with a diagnosis of IgAV. [15]

None of the patients showed superimposed petechiae and purpura over the thoracolumbar spine. All but one of the ten patients eventually developed a rash, which appeared one day after admission in patient 3. [3] Patient 8 never developed a purpuric rash, but decreased plasma factor XIII activity strongly suggested a presumptive diagnosis of IgAV. [8] Plasma factor XIII activity was decreased in four out of five patients measured. [4,7,8,9] Decreased plasma factor XIII activity in IgAV may result from leukocyte protease degradation or consumption around affected vessels. [16] Decreased plasma factor XIII activity was not only a useful diagnostic clue for IgAV prior to rash development, but also closely associated with abdominal pain. [8,16] Our patient had normal plasma factor XIII activity, possibly due to mild disease or lack of abdominal pain.

Second, PLSE caused sudden onset of severe back pain. Seven patients went directly to the emergency room, and nine refused to walk because of severe lumbar pain. Our patient was transferred by ambulance, because any backward movement caused him to scream. Refusal to walk was a common symptom in children with IgAV who had arthralgia in the ankle and knee joints. [9] IgAV may not be a primary consideration for gait disturbance associated with low back pain. [9] The precise mechanism of PLSE was not fully understood and remained unclear. PLSE was associated with obesity in adults. [17] The patient and other patients had no obesity, suggesting that obesity related PLSE is unlikely in children. The patient also had no cardiac, hepatic, or renal dysfunction causing edema. Symptoms of IgAV generally appeared with the direction of gravity. [18] PLSE was often seen in hospitalized patients with prolonged bed rest. [19] The patient has not been in a situation where gravity has been applied to the back.

Ultrasound images of the swollen posterior lumbar region showed minimal soft tissue imbibition. [8] Magnetic resonance imaging and computed tomography scans have been reported to show edema in the lumbosacral subcutaneous facial planes without destruction of the underlying structures, which is different from a typical hematoma or abscess. [1,3,4,5,7,9] Nakajima et al. reported that the underlying mechanisms of PLSE may be lymphatic

or interstitial fluid accumulation due to disturbance of the lumbar fascia. [17] IgAV may affect the lumbar region. Capillary endothelial IgA deposition caused an increase in capillary permeability and leakage from blood vessels into the surrounding tissue, resulting in subcutaneous edema. [5,8,19] The transient PLSE may result from the short-term production/development and subsequent rapid clearance of IgA, followed by rapid mobilization of excess subcutaneous fluid by lymphatic drainage. [5] Hypertension may also contribute to the increase in intravascular pressure, but our patient did not have hypertension. [20]

Rapid accumulation in the soft tissue spaces must cause severe lumbar pain and swelling. [1,3,4,5,6,7,8,9] Corticosteroids were given to four patients, three of whom had abdominal pain. [2,4,9] The remaining five patients received non-steroidal anti-inflammatory drugs. [1,3,5,6,8] One patient received antihistamines and the rest received nothing. [7,9] The lumbar pain and swelling completely resolved within six days without recurrence.

4. Conclusion

IgAV may rarely involve the lumbar region and cause sudden onset of atraumatic pain and swelling in the lumbar spine. PLSE was a self-limiting symptom of limited duration and may not require specific treatment such as corticosteroids.

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None

State of Competing Interests

The authors have no competing interest.

List of abbreviations

IgAV: IgA vasculitis, PLSE: posterior lumbar subcutaneous edema, ACE: angiotensin converting enzyme, HAE: hereditary angioedema.

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