Deep Vein Thrombosis Related with Henoch-Schönlein Purpura in Pediatric Patient

Sirada Srithanetsakul MD, Burawat Nilrod MD.
Phramongkutklao Hospital, Bangkok, Thailand.

ABSTRACT:
Henoch-Schönlein Purpura is a condition within the vasculitis groups. It can result in manifestations, including abdominal discomfort, kidney problems, joint inflammation, and palpable purpura. In some instances, it may even lead to the development of thrombosis. An illustrative case involved a boy diagnosed with Henoch-Schönlein Purpura who experienced vein thrombosis in both legs. The patient experienced an improvement in his symptoms after receiving enoxaparin and colchicine.
Key words: Henoch-Schönlein Purpura, Deep vein thrombosis

Introduction

Thrombosis is a rare but fatal outcome, even though prothrombotic conditions are frequently associated with Henoch-Schönlein Purpura (HSP)[1]. To improve outcomes, it is crucial to recognize life-threatening problems early in high-risk patients. We reported on an 11-year-old boy with HSP who had deep vein thrombosis along both legs.

Case report

An 11-year-old child, who was previously healthy, was brought to our hospital complaining of arthralgia for 2 days. He had a purpuric rash on his lower leg and arthralgia in his left knee for six weeks. He reported that he had arthralgia in both ankles, a purpuric rash that worsened, and lower leg edema for two days.

On general examination, his lower legs were edematous; the circumferences of the right and left lower legs were 41 and 36 centimeters, respectively. Both lower legs had multiple symmetrical palpable purpuric papules and plaques. (Figure 1) No mucosal involvement was seen. Other physical examinations were unremarkable.

Corresponding author: Burawat Nilrod MD., Email: nanurz_stn@docchula.com

Received: 12 December 2023 Revised: 17 May 2024 Accepted: 12 August 2024

The initial laboratory tests revealed the following: hemoglobin 10.7 g/dL, leukocyte count: 5.7 x 10^9/L with 56.2% polymorphonuclear leukocytes, 36.4% lymphocytes, and platelet count: 360/mm^3. The results of the serum coagulogram, blood urea nitrogen, creatinine, liver function test, erythrocyte sedimentation rate, C-reactive protein, and urine analysis were all within their normal limits. Anti-nuclear antibody, anti-dsDNA, and anti-neutrophil cytoplasm antibodies (ANCA) were negative. The complement concentration was normal. The anti-HIV test was also negative.

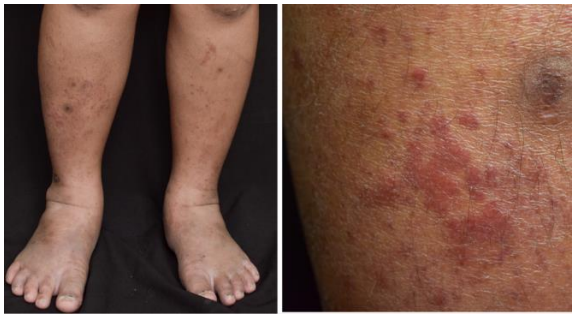

Figure 1 Palpable purpura on lower legs

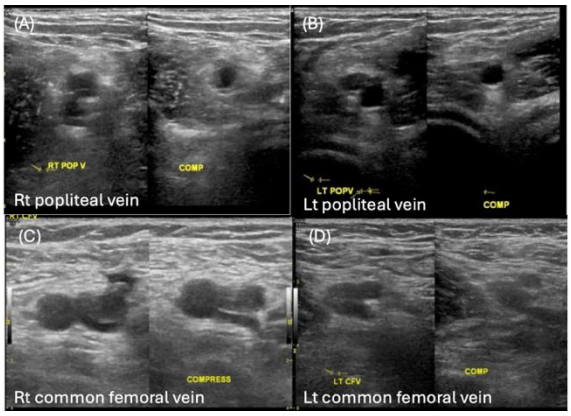

Figure 2A-D A venous doppler ultrasound of both lower extremities demonstrates enlarged and non-compressible veins of right popliteal vein, left popliteal vein, right common femoral vein and left common femoral vein respectively.

A hypercoagulability test revealed elevated homocysteine, fibrin degradation products (D-dimer), and antithrombin III levels. Factor VIII, protein C, protein S, and serum fibrinogen were all within the normal range. The results for antiphospholipid antibodies (anti-cardiolipin, anti-beta2-glycoprotein, and lupus anticoagulant) were negative.

A venous doppler ultrasound of the lower extremities revealed vein thrombosis in the extremities specifically in the bilateral popliteal veins, common femoral veins as well as femoral veins. (Figure 2)

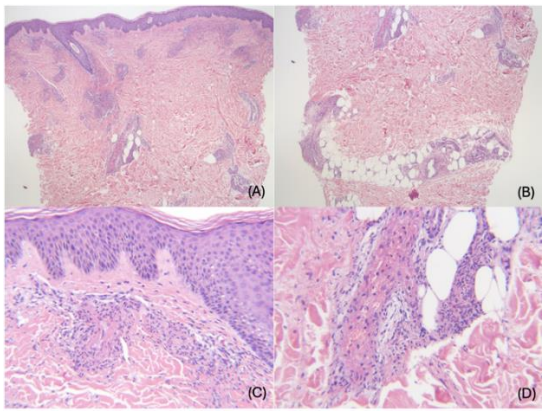

Figure 3A-D Skin biopsy of the left lower leg showed dense superficial and mid-perivascular infiltration consisting of a mixed infiltrate of inflammatory cells composed of neutrophils, nuclear dust, and extravasated erythrocytes, some of the small blood vessels showed deposition of eosinophilic fibrinoid material in and around the vessel wall. (Figure 3A-D: H&E, 40x, 40x, 200x and 200x, respectively)

Skin biopsy of the left lower leg showed dense superficial and mid-perivascular infiltration consisting of a mixed infiltrate of inflammatory cells composed of neutrophils, nuclear dust, and extravasated erythrocytes, some of the small blood vessels showed deposition of eosinophilic fibrinoid material in and around the vessel wall. (Figure 3)

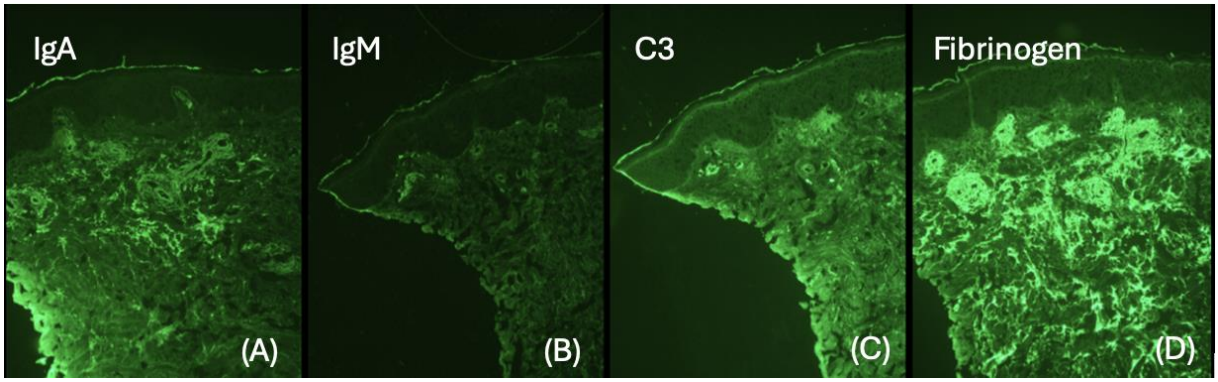

Figure 4A-D Direct immunofluorescence showed conspicuous granular deposits of IgA, IgM, and C3 in the perivascular vessels. (Figure 4A-D: 100x, IgA, IgM, C3 and fibrinogen, respectively)

Direct immunofluorescence showed conspicuous predominated granular deposits of IgA at perivascular vessels in upper dermis. IgM, and C3 were also presented in the perivascular vessels in weaker intensity. (Figure 4)

According to the clinical symptoms and test results, deep vein thrombosis and HSP were identified as the patient's conditions. During his treatment, he received enoxaparin (1mg/kg q12hr SQ) and colchicine (0.6mg) 1 tablet orally twice daily. During the follow-up period of one month, there was improvement, in both symptoms and signs of HSP and doppler venography did not find any evidence of blood clotting.

Discussion

Patients with autoimmune conditions including SLE, rheumatoid arthritis and Sjogren's syndrome have reportedly had blood clotting issues. This condition has also been linked to systemic vasculitis such Behcet's disease, ANCA-associated vasculitis, and large vessel vasculitis[1]. Venous and arterial thrombosis is an uncommon side effect of HSP. Table 1 displays the 13 cases of thrombosis linked to HSP that Dhaliwal[2], Xu YY[3] and Baylon AK[4] described.

Table 1 Reported cases of thrombosis related to Henoch-Schönlein Purpura

Author Reference	Sex	Age	Location of thrombosis	Treatment	Outcome	Associated factors
Luan Li[1]	M	14	Superior mesenteric vein	Steroids, heparin	Recovered with no relapse after 2 months follow-up	-
Dhaliwal K[2]	F	15	Left distal basilic and cephalic veins	Steroids, heparin	Recovered	-
			Ileal ischemia	IVIG, warfarin, IV cyclophosphamide	Recovered with no relapse after 6 months follow-up	
Abend NS[5]	M	15	Superior sagittal sinus, straight sinus, and transverse sinuses	Steroids, heparin, warfarin	Recovered with no relapse after 3 months follow-up	Abnormal lupus anticoagulant

Table 1 Reported cases of thrombosis related to Henoch-Schönlein Purpura

Author Reference	Sex	Age	Location of thrombosis	Treatment	Outcome	Associated factors
Topaloglu R[6]	M	15	Iliac and femoral vein	Steroids, heparin	Recovered	High homocysteine and factor VIII level
Sari I[7]	M	37	Bilateral common femoral veins	Steroids, anticoagulant	Recovered	Celecoxib (COX-2 inhibitor)
			Penile dorsal vein	Intracarvernous SK irrigation	Recovered with no relapse after 3 years follow-up	
Xu YY[3]	M	12	Pulmonary embolism and left leg vein	Heparin, urokinase Steroids	Recovered	-
Diana A[8]	M	8	Spermatic vein	Steroids, heparin	Recovered	-
Lind J[9]	M	9	Penis	Caudal anesthesia Aspiration of corpus carvernosa and irrigation with saline	Recovered	-
Park YW[10]	M	29	Iliac vein, inferior vena cava and femoral vein	Steroids, heparin, warfarin	Recovered	-
Baylon AK[4]	F	14	Superior sagittal sinus, transverse sinus, common iliac vein	Enoxaparin	Recovered	-
Abebe M[11]	M	58	Portal vein thrombosis	Steroids, bowel resection	Death	-
Monastiri K[12]	M	6	Tibial and fibular artery	Heparin	Recovered with no relapse after 4 years follow-up	High titer of Anticardiolipin IgG and IgM
Canpolat U[13]	M	33	Right coronary artery	Drug-eluting stent placement and percutaneous coronary intervention	Recovered with no relapse after 6 months follow-up	-

HSP has been observed to have an association with thrombosis in males compared to females. Most cases were venous thromboses. Arterial thrombosis has only been reported twice; one involved the coronary artery, the other the tibial and fibular arteries[2]. Thrombosis in HSP is increasingly reported but is still rare.

People with this condition generally respond positively to treatment involving steroids and anticoagulants. However, there was one case where an individual succumbed to an illness that affected portal vein. It was also discovered that patients who had high levels of homocysteine and factor VIII, also, the presence of antiphospholipid antibodies has

been related to higher risk of thrombosis in HSP[2].

In our case, we illustrated that venous thrombosis of the common femoral, femoral, and popliteal veins was associated with HSP and high homocysteine levels in an 11-year-old male. Neither antiphospholipid syndrome screening nor factor VIII was abnormal in this case. After receiving treatments, the signs, and symptoms of HSP improved. Thrombosis could not be detected by doppler venography.

The mechanism of the hypercoagulable state is still incompletely understood. It is assumed that impaired activation of the fibrinolytic system and an activated coagulation system play an important role in the hypercoagulable state in HSP[1]. Elevated factor VIII, homocysteine, lipoprotein A, von Willebrand factor, and antiphospholipid antibodies are associated with the prothrombotic state in HSP[2]. These factors together with the inflammatory state in HSP vasculitis increase the risk of thrombosis.

Conflict of Interest and Financial Support

The research was conducted independently without any backing, from corporations or other funding sources.

References

1. Li L, Zhang J, Zhang Y, Ji H. Thrombosis warning in children suffering from henoch-schonlein purpura. Indian J Dermatol 2013;58:409.
2. Dhaliwal KK, Lile NA, Tan CL, Lim CH. Life-threatening complications of Henoch-Schönlein purpura: diffuse alveolar haemorrhage, venous thrombosis and bowel ischaemia. BMJ Case Rep 2020;13:e235905.
3. Xu YY, Huang XB, Wang YG, Zheng LY, Li M, Dai Y, Zhao S. Development of Henoch-Schoenlein purpura in a child with idiopathic hypereosinophilia syndrome with multiple thrombotic onset: A case report. World J Clin Cases 2023;11:952–61.
4. Baylon AK, Tan HP, Naranjo ML, et al. A Rare Case of Thrombotic Storm in Henoch-Schonlein Purpura. Res Pract Thromb Haemost 2021;5:P593.
5. bend NS, Licht DJ, Spencer CH. Lupus anticoagulant and thrombosis following Henoch-Schonlein purpura. Pediatr Neurol 2007;36:345-7.
6. Topaloglu R, Bayrakci US, Cil B, Orhon D, Bakkaloglu A. Henoch-Schonlein purpura with high factor VIII levels and deep venous thrombosis: an association or coincidence? Rheumatol Int 2008;28:935–7.
7. Sari I, Akar S, Secil M, et al. Thrombosis and priapism in a patient with Henoch-Schonlein purpura. Rheumatol Int. 2005;25:472–4.
8. Diana A, Gaze H, Laubscher B, De Meuron G, Tschantz P. A case of pediatric Henoch-Schönlein purpura and thrombosis of spermatic veins. J Pediatr Surg 2000;35:1843.
9. Lind J, Mackay A, Withers SJ. Henoch-Schönlein purpura and priapism. J Paediatr Child Health 2002;38:526–7.
10. Park YW, Park JJ, Lee JB, Lee SS. Development of Henoch-Schönlein purpura in a patient with Behçet's disease presenting with recurrent deep vein thrombosis. Clin Exp Rheumatol 2007;25:S96-8.
11. Abebe M, Patnaik A, Miller F, et al. Intestinal infarction and portal vein thrombosis in a patient with Henoch Schonlein Purpura. Case Rep Rheumatol 2012;2012:1-4.
12. Monastiri K, Selmi H, Tabarki B, Yacoub M, Mahjoub T, Essoussi AS. Primary antiphospholipid syndrome presenting as complicated Henoch-Schönlein purpura. Arch Dis Child 2002;86:132–3.
13. Canpolat U, Yorgun H, Sahiner L, Kabakçi G. Myocardial infarction due to coronary thrombosis in a patient with Henoch-Schönlein purpura. Herz 2012;37:801-3.