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Effort Thrombosis (Paget-Schroetter Syndrome) in a 16-Year-Old Male

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Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Male, 16
Final Diagnosis: Effort thrombosis (Paget-Schroetter Syndrome)
Symptoms: Swollen arms
Medication: —
Clinical Procedure: —
Specialty: Metabolic Disorders and Diabetics

Objective: Rare disease





Background: Thrombotic events in otherwise healthy pediatric patients are rare. In patients presenting with limb swelling, thrombosis must be considered in the differential diagnosis. In pediatric patients with thrombosis, there has been wide variability in the rates of associated thrombophilia. Many pediatric patients may instead have other contributors such as venous catheters or physical activity.

Case Report: We present a case of bilateral upper extremity deep venous thrombi in a previously healthy 16-year-old male. The patient presented with swelling and pain in both arms after several days of weight-bearing exercise. Following emergency department evaluation with ultrasound and laboratory testing, the patient was diagnosed with effort thrombosis – also known as Paget-Schroetter syndrome – and rhabdomyolysis.

Conclusions: This case of Paget-Schroetter syndrome is distinguished by elevation in creatine kinase and transaminases. While these findings can be due to physical exertion and effort, effort thrombosis is not classically associated with laboratory abnormalities except an elevated D-dimer. The significance of these laboratory test result abnormalities remains unclear. Given the rarity of effort thrombosis, further epidemiological study is warranted to determine if these laboratory findings are seen in other cases, and, if so, what implications they may have for management and prognosis.

MeSH Keywords: Upper Extremity Deep Vein Thrombosis • Hematology • Rhabdomyolysis • Transaminases • Pediatrics

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Background

Venous thromboembolism (VTE) is a condition that includes deep vein thrombosis (DVT) and pulmonary embolism (PE). DVT, which occurs primarily in the lower extremities, constitutes about two-thirds of VTE and is postulated to result from “Virchow’s triad” of stasis, hypercoagulability, and endothelial injury.

Symptomatic VTE is rare in children, affecting only about 0.7 per 100 000 children [1]. In the population as a whole, the overall incidence of first-time VTE is 100 persons per 100 000 annually, and occurrence of the condition rises substantially with increasing age [2]. VTE in pediatric patients is often associated with central venous catheters, while studies have shown wide variability in the prevalence of inherited thrombophilia (13–79%) in children with VTE [3–6]. In contrast to PE in adults, PE in children is less prone to recurrence and is rarely fatal [1].

Case Report

A previously healthy 16-year-old male presented to the emergency department (ED) with a chief complaint of swollen arms. Two weeks prior to presentation, he had initiated a 5-day regimen of strenuous upper extremity weightlifting, focusing on bicep curls. He had no other unusual physical activity and no reported injury. Two days before presentation, he developed bilateral arm swelling with some associated pain. His primary physician found a creatine kinase (CK) level of 6540 IU/L, prompting referral to the ED for further evaluation.

The patient had no history of similar episodes. There was no family history of thrombophilia and no recent travel or surgery. He denied use of supplements or steroids.

The patient’s physical examination revealed bilateral swelling in the triceps, elbows, and proximal third of the forearms bilaterally, with associated erythema. He had full range of motion of his joints, and no tenderness. Radial and ulnar pulses, strength, and sensation were intact. There was no lower extremity edema. The other examination results were unremarkable.

Evaluation in the ED included laboratory tests (complete blood count, comprehensive metabolic panel including liver function tests, creatine kinase, erythrocyte sedimentation rate, c-reactive protein, and urinalysis), chest x-ray, and bilateral upper extremity venous duplex ultrasound. It also included. Intravenous access was established and a bolus of crystalloid fluids administered.

Abnormal laboratory results included CK of 4385 IU/L, aspartate transaminase (AST) of 163 IU/L, and alanine transaminase

(ALT) of 90 IU/L. Upper extremity ultrasound demonstrated non-compressibility of the right cephalic and left brachial veins, without color flow, indicating bilateral thrombus.

A heparin drip was initiated for treatment of DVT, and the patient was admitted to the pediatric department. During his admission, he was evaluated by the hematology department. No inherited thrombophilia was identified, and he was given a diagnosis of effort-related deep venous thrombosis – also known as Paget-Schroetter syndrome.

In the hospital, intravenous fluid administration was continued. His heparin drip was discontinued, and he was subsequently started on enoxaparin. His laboratory values progressively improved, and on the day of discharge his CK was 1125 IU/L, AST was 74 IU/L, and ALT was 72 IU/L. His arm discomfort and swelling also improved significantly prior to discharge. The patient was subsequently lost to follow-up.

Discussion

Primary DVT of the upper extremity has an annual incidence estimated at 1–2 cases per 100 000 [7,8] and constitutes 1–4% of all episodes of DVT [9]. Diagnosis is typically made by ultrasound, although the criterion standard is contrast venography [10,11]. Paget-Schroetter syndrome most commonly affects the axillary and subclavian veins [12,13]. Thoracic outlet abnormalities or repetitive activity causing trauma to the venous endothelium are common predisposing factors [12].

Paget-Schroetter syndrome most commonly presents in athletic males during the 4th decade of life [14–16]. Physical examination demonstrates limb edema, often with distal cyanosis but intact pulses [8,17–19]. Possible early complications include pulmonary embolism and secondary arterial occlusion, and post-thrombophlebitic syndrome may develop later [9,16].

Results of laboratory studies in Paget-Schroetter syndrome are generally normal, aside from an elevated level of d-dimer [20]. Our case is notable for elevations of CK, AST, and ALT. Given a serum half-life of 1.5 days for CK [21], it seems unlikely that the CK elevation is attributable to an exercise regimen discontinued 9 days earlier. Although rhabdomyolysis can be caused by localized DVT [22,23] (or, alternatively, can mimic it [24]), this appears to be highly unusual with Paget-Schroetter syndrome, as we could identify only 1 such case reported in the literature [25].

We found no prior reports of elevated transaminases with Paget-Schroetter syndrome. Our patient had no previous transaminase results on record for comparison and was subsequently lost to follow-up. However, he had no known history

of abnormal values or congenital muscular disorders. As noted, his transaminase levels progressively and significantly declined as his symptoms improved. Although exercise can produce an elevation of transaminases [26], this would again seem an unlikely cause considering the duration of time since his exercise regimen had stopped. Assuming that the patient did not have some coincidental underlying condition that could not be confirmed due to the lack of follow-up, the timing suggests a direct relationship between the elevated transaminase levels and the effort thrombosis.

No consensus exists for managing Paget-Schroetter syndrome. Strategies may include combinations of anticoagulation or fibrinolytics. Surgical interventions to decompress the thoracic outlet may also be considered, and some evidence suggests that nonsurgical management may be associated with poorer long-term outcomes [15,27–32]. Newer evidence has emphasized a more aggressive approach using catheter-directed thrombolysis, but this approach is not uniform, and sources such as UpToDate still summarize the evidence by suggesting that “patients who are diagnosed with primary upper extremity deep vein thrombosis are initially managed with measures to improve their comfort and are anticoagulated. ... [the] decision to proceed with thrombolysis or thoracic outlet decompression is based upon symptom severity and the type of

associated anatomic abnormality” [33]. While it is appropriate to strongly consider these more aggressive interventions when managing Paget-Schroetter syndrome, these were deferred in our case due to the degree of symptoms and initial clinical improvement with the anticoagulation recommended by our hematology department.

Conclusions

Paget-Schroetter syndrome is rarely seen in pediatric or adolescent patients but should be considered when upper extremity swelling and pain follows exertion. Although there is no definitive diagnostic test, Paget-Schroetter syndrome is suggested by findings of DVT in the appropriate clinical context. Our case, while typical in terms of history and physical findings, is unusual given the associated laboratory test results showing elevated CK and transaminases. Patients with this condition require admission to the hospital for evaluation by a hematology specialist and appropriate treatment of DVT.

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