CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 49 (2018) 81–86

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Paget-Schroetter syndrome as a result of 1st rib stress fracture due to gym activity presenting with Urschel's sign – A case report and review of literature

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ARTICLE INFO

Article history:
Received 10 April 2018
Accepted 15 May 2018
Available online 20 June 2018

Keywords:
Deep vein thrombosis
Paget-schrotter syndrome
Axillary-subclavian venous thrombosis
Pulmonary embolism
Thrombosis
Upper extremity deep vein thrombosis

ABSTRACT

BACKGROUND: Paget-Schrotter Syndrome (PSS) also known as ëffort thrombosisïs a form of primary thrombosis in the subclavian vein at the costoclavicular junction is usually seen in younger patients after repeated strenuous activity of the shoulders and arms. When occurring in younger patients, PSS presents itself with predisposing factors such as unilateral dull, aching pain in the shoulder or axilla and swelling of the arm and hand.

CASE PRESENTATION: We report a rare case of unusual left axillo-subclavian vein thrombosis following narrowing of thoracic outlet due to stress injury of rib fracture likely during gym activity in absence of other clear risk factors and a negative hypercoagulable workup in a 27-year-old man who was admitted as left deltoid rupture 5 days after his usual gym. Subsequently he was transferred under vascular surgery for thrombectomy.

CONCLUSION: In addition to the unusual location in the left upper extremity in our case, the absence of common etiologic factors makes our case of Paget-Schroetter Syndrome a very unique one. This case report aims to discuss the common causes of PSS in order to raise a high index of suspicion in certain groups of patients. This will allow early identification and avoidance of catastrophic outcomes such as pulmonary embolism and stroke.

1. Introduction

Spontaneous effort-induced upper extremity deep vein thrombosis, also known as Paget-Schroetter syndrome (PSS), is a rare phenomenon first described by Cruveilhier in 1816, with the first elaborate account provided by James Paget in 1875 [1,2].

This case report aims to discuss the common causes of PSS in order to raise a high index of suspicion in certain groups of patients. This will allow early identification and avoidance of catastrophic outcomes such as pulmonary embolism and stroke. Please ensure you state in your introduction that the work has been reported in line with the SCARE criteria [19].

2. Case report

A 27 year old male patient presented with left upper limb pain and swelling, following inclined press exercises at the gym 5 days prior. He visited the gym regularly 3 times a week for the last 6 months, predominantly doing bodybuilding exercises using the inclined press machine.

At the Emergency Department, X-ray of the left shoulder was done showing no fracture or dislocation. A provisional clinical diagnosis of anterior deltoid muscle tear was made, he was treated with analgesia and given an orthopaedic outpatient appointment. The pain and swelling persisted, hence the patient consulted a general practitioner who referred him to our Emergency Department. He was admitted to the Department of Orthopaedic Surgery with the impression of anterior deltoid muscle rupture.

During our inpatient review, we noted a diffuse swelling of the left upper arm and forearm. There were dilated veins and bruising over the anterior shoulder and the entire upper arm (Urschel's sign) (Picture 1).

His x-ray revealed healed fracture left 1st and 2nd rib and a MRI was arranged, showing subclavian vein thrombosis with an old fracture and callus formation at the left 1st rib (Pictures 2 and 3). A subsequent venous Doppler ultrasound was done, which revealed venous thrombosis extending from the proximal subclavian vein to the basilic vein (Picture 4). He was referred to the Vascular Surgeons and Computed Tomography (CT) angiography was done

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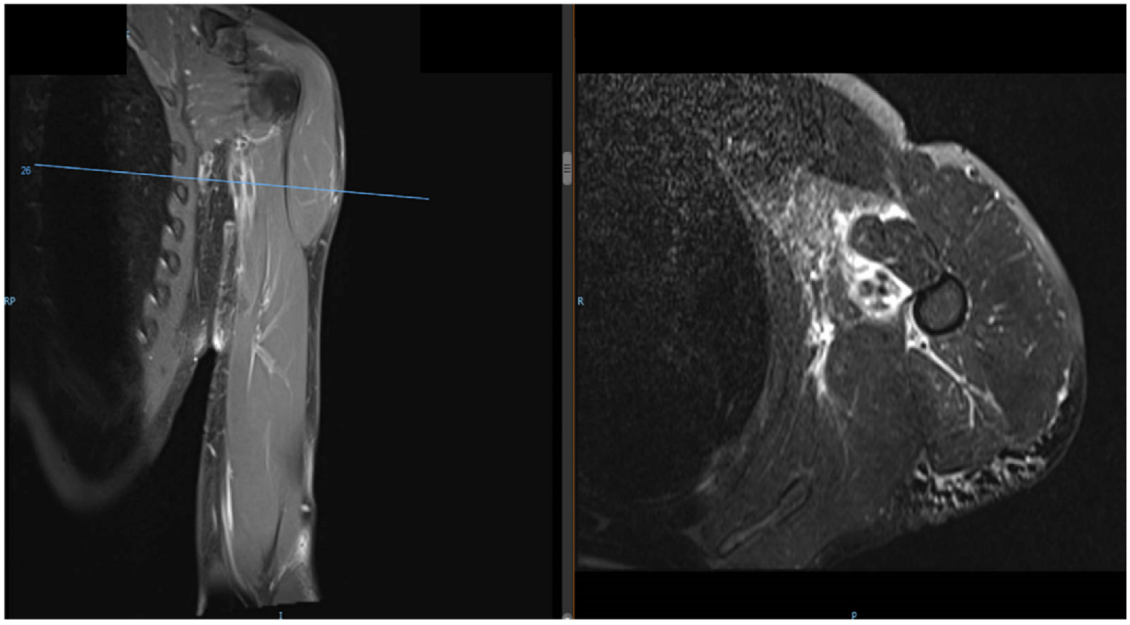
https://doi.org/10.1016/j.ijscr.2018.05.029

Picture 1. Urschel's sign.

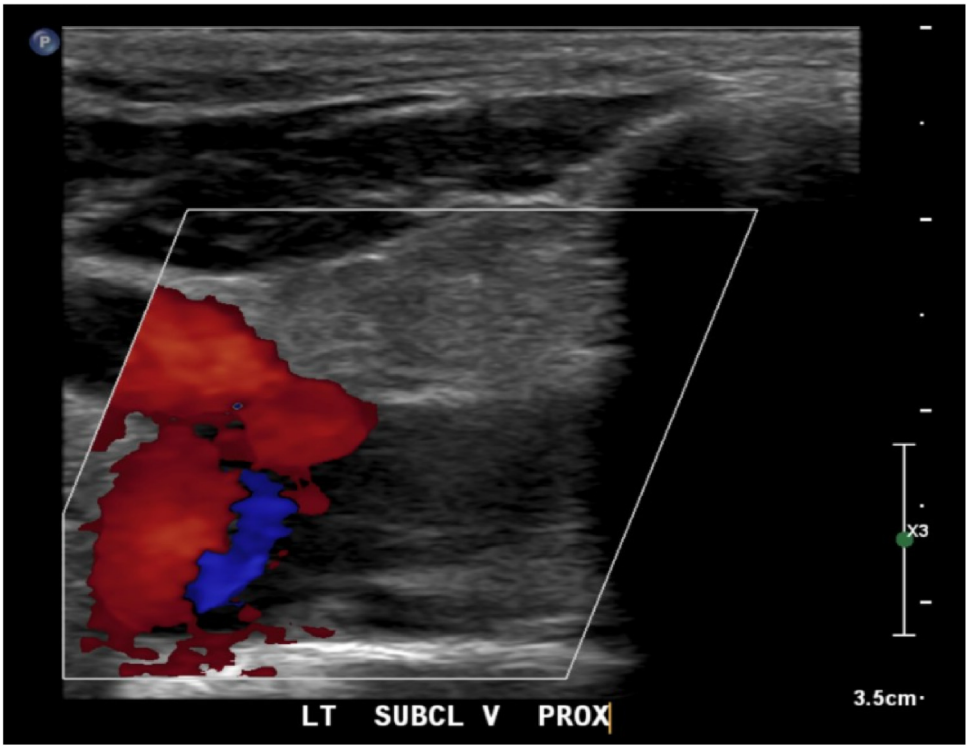
picture 2. xray-left 1st and 2nd rib healed fracture with callus.

which reported healed fractures of the left 1st to 5th fibs. The left 1st rib fracture demonstrated exuberant callus, which compressed the left subclavian vein against the left clavicle (Picture 5). The patient was started on a therapeutic dose of Enoxaparin Sodium subcutaneous injection and transferred to the Department of Vascular Surgery for thrombectomy.

6 days after angiojet thrombectomy of the left subclavian vein, the patient was discharged from the hospital with oral anticoagulant Rivaroxaban. At 1-month follow-up, his left arm symptoms had completely resolved. He was back to normal activity and work but was advised to refrain from gym exercises that precipitated PSS. Option of left 1st rib excision was discussed but he required more time to consider.



Picture 3. MRI showing left subclavian vein thrombosis and old fracture of the left 1st rib.



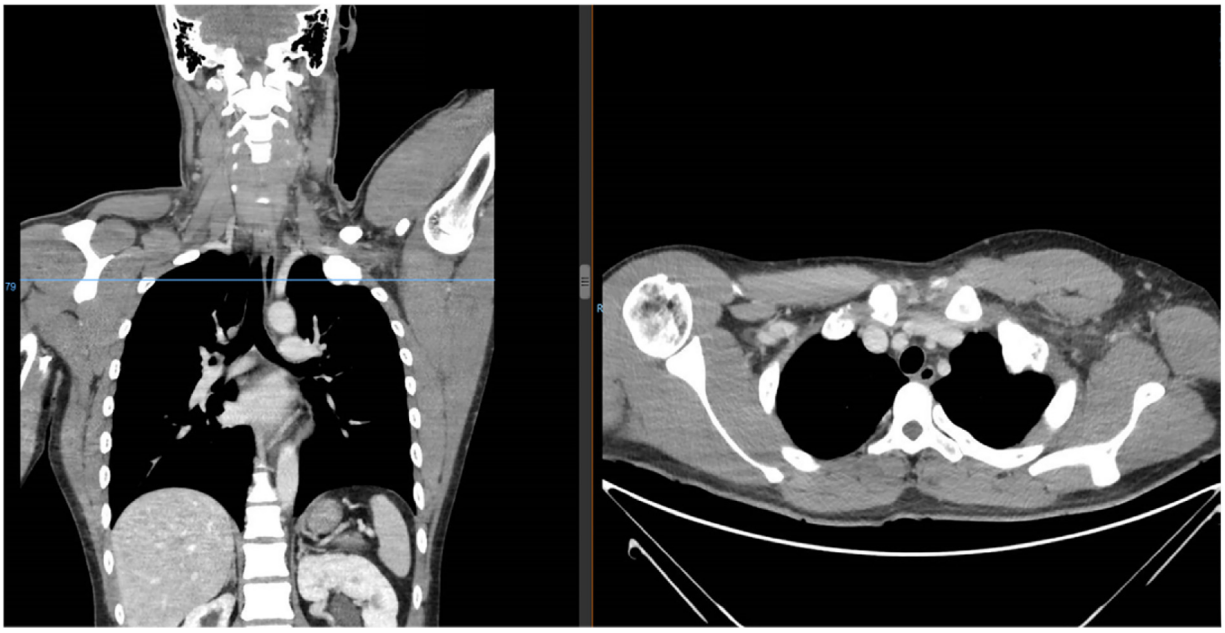
Picture 4. Venous doppler ultrasound showing venous thrombosis.

3. Discussion

Upper extremity deep vein thrombosis (UEDVT) is reported mainly in adults and 80% of cases are associated with indwelling central venous catheters, pacemakers or defibrillator leads placed in the upper limb venous system or with other secondary causes such as malignant disease, surgery or trauma to the upper limb, pregnancy, use of oral contraceptives or ovarian hyper stimulation syndrome [2].

Effort induced UEDVT is a rare condition precipitated by mechanical exertion involving the upper limb (PSS) with or without narrowing of thoracic outlet leading to external compression on the subclavian vein.

On literature review, majority of case reports show an association between PSS and activities that involve repetitive or prolonged hyper-abduction or external rotation of the shoulder joint- particularly in the overhead position which includes overhead lifting or throwing (Table 1).



Picture 5. CT images showing excuberant callus of the left first rib, compressing the left subclavian vein against the left clavicle.

Table 1
Case reports of PSS and associated physical activity.

No	Author	Time of Publication	Age/Gender	Side Involved	Physical Activity	Delay in Presentation	Management
1	Yagi S [11]	Jul-13	31/Male	Left	Baseball (pitching)	1 day	Anticoagulation
2	Sancho-González [12]	May-17	38/Male	Left	Triathlon	2 days	Anticoagulation
3	Sanson H [13]	Dec-16	23/Female	Right	Violin playing	1 day	Anticoagulation
4	Edo Fleta G [14]	Jun-16	38/Female	Right	Swimming	1 day	Percutaneous transluminal angioplasty
5	Pekić P [15]	Apr-16	28/Male	Left	Carpentry (pneumatic drill)	1 day	Anticoagulation
6	Ijaopo R [16]	Mar-16	37/Male	Right	Judo	7 days	Catheter directed thrombolysis
7	Thiruchelvam N [17]	Oct-15	43/Male	Left	Weight lifting	14 days	Catheter directed thrombolysis
8	Lutter C (2 cases) [18]	Oct-15	41/Male	Right	Roof climbing	1 day	Anticoagulation
			34/Female	Left	Rock climbing	7 days	Anticoagulant and staged rib resection
9	Meena	Oct-15	40/Male	Right	Manual labour	5 days	Anticoagulation
10	Shimada T	Mar-15	17/Male	Left	Athlete (not specified)	7 days	Anticoagulation
11	Bullock C	Aug-16	23/Male	Right	Military	1 day	Anticoagulant and staged rib resection
12	Beasley R	Jun-15	40/Male	Right	Photography (cameraman)	21 days	Catheter directed thrombolysis
13	Kondo T	Jul-15	65/Male	Right	Clerical work	2 days	Anticoagulation
14	Stein CM	Sep-15	45/Female	Left	Furniture lifting	5 days	Catheter directed thrombolysis and staged rib resection
15	Jackson SS	May-14	19/Male	Right	Baseball (pitching)	21 days	Anticoagulation
16	Keene DJ	Apr-15	28/Female	Right	Surfing	3 days	Catheter directed thrombolysis
17	Spencer TR	Aug-14	16/Male	Bilateral	Weight lifting	2 days	Anticoagulation
18	Kellar J	Jul-14	19/Male	Right	Weight lifting	2 days	Catheter directed thrombolysis
19	Young K	May-14	9/Male	Left	Horse riding	1 day	Catheter induced thrombolysis
20	Rainey CE	Apr-14	34/Male	Right	Military	4 days	Balloon angioplasty and staged rib resection
21	Dep A	Jul-13	18/Male	Right	Canoeing	2 days	Catheter directed thrombolysis
22	Aguilar Shea AL	Mar-13	41/Male	Right	Spontaneous	Few weeks	Anticoagulation
23	Drakos N	Mar-13	29/Male	Right	Waiting (waiter)	3 days	Catheter induced thrombolysis
24	Sayın A	Jun-12	52/Male	Right	Spinning exercise	5 days	Anticoagulation
25	Allana AM	Mar-11	32/Male	Right	Triathlon	3 days	Anticoagulation
26	Girma F	Jan-10	25/Male	Left	Heavy lifting (TV)	3 days	Anticoagulation
27	Pysklywec M	Mar-11	38/Male	Left	Industrial mechanic work	1 day	Catheter directed thrombolysis
28	Singh AP	Aug-09	27/Male	Left	Previous shoulder soft tissue injury	3 days	Anticoagulation
29	Phipps C	Jun-10	33/Male	Right	Video gaming	14 days	Anticoagulation
30	Toya N	Dec-12	23/Female	Right	Push ups	5 days	Anticoagulation

Our patient developed his symptoms following bench press activity, with a background of similar exercises for the past 6 months. This suggests repetitive overhead activity as possible causative factor for PSS. However, pathogenesis of PSS likely involves not only repetitive damage to the vein's endothelium, but also anatomic variants associated with thoracic outlet syndrome [3,4],.This can be postulated from the low prevalence of PSS in weight lifters, despite their repetitive heavy weight training. Those anatomic variants could decrease normal mobility of the subclavian vein, hence increasing its susceptibility to injury from arm

activity. Repetitive endothelial damage results in vascular intimal hyperplasia, inflammation, fibrosis, and activation of the coagulation cascade, leading to thrombus formation [5,6]. Toya N, et al described the first case of exercise–induced thrombosis in a young woman following repetitive push-ups [7]. Push-up exercises can cause serratus anterior

hypertrophy, potentially leading to compression and micro trauma of the subclavian vein. In our patient, compression of the subclavian vein may have occurred due to a combination of weight training and stress fracture of first rib leading to narrowing of the thoracic outlet which triggered the coagulation cascade (on non-dominant side of limb) and extensive thrombus formation.

Differential diagnoses to be considered in a patient with upper limb pain and swelling include cellulitis, lymphedema, traumatic muscle injury, neoplastic venous compression, and superficial vein thrombosis. However, presence of dilated collateral veins around the shoulder, also known as Urschel's Sign, is characteristic of PSS and should raise clinical suspicion of a vascular etiology [8]. This is critical in preventing associated complications such as pulmonary embolism (reported to be 7–20%), superior vena cava syndrome, stroke, and compartment syndrome [9]. In our case, diagnosis was delayed by 6 days as the patient was initially discharged with the provisional diagnosis of left anterior deltoid muscle tear.

Further evaluation needs to be done in the setting of upper limb pain and swelling. Detailed imaging should include Doppler Ultrasound in the initial work up as it is non-invasive with high sensitivity and specificity. MRI and Contrast Venography may also be considered to aid diagnosis. Laboratory investigations should include complete blood counts (with attention to platelet counts in order to exclude other etiologies), as well as a complete pro-thrombotic work up to exclude secondary causes of venous thrombosis [6]. Diagnosis of deep vein thrombosis in our patient was confirmed with MRI and venous Doppler Ultrasound. A hypercoagulable state was unlikely as initial pro-thrombotic screen was negative.

PSS if diagnosed early, allows early institution of medical and surgical treatment, with good resultant outcome and prognosis. Conventional treatment usually involves anticoagulation and thrombolysis. Skeletal abnormality causing thoracic outlet and hence venous obstruction should also be addressed. Our patient was referred to the vascular surgeons for definitive treatment. Physiotherapy and rehabilitation should also be included as part of patient's management, especially with muscular causes of venous blood flow obstruction [10].

4. Conclusion

Although uncommon, PSS may result in life-threatening sequelae such as pulmonary embolism. As such, there should be a high index of suspicion for PSS in any case of upper limb swelling with a preceding history of repetitive exercises. Laboratory investigations should include complete blood counts and complete pro-thrombotic work up to exclude secondary causes. Detailed imaging such as Doppler Ultrasound, MRI and Contrast Venography should be considered. It is often difficult to differentiate between a muscle rupture and PSS, hence an important finding to look for is the presence of dilated superficial veins (Urschel's sign). A brief screen for any thoracic outlet syndrome on physical examination will also be helpful in determining causative factors for PSS. PSS if diagnosed and treated early, usually results in excellent outcome and prognosis.

Conflicts of interest

None of the authors have conflict of interest.

Sources of funding

No source of funding is involved in the collection, analysis and interpretation of data; in the writing of the manuscript; and in the decision to submit the manuscript for publication.

Ethical approval

Study is a case report without any added intervention hence exempted for ethical approval from research board of institution.

Consent

Informed consent taken before writing this report.
have patient consent for the publication of this case report.

Author contribution

Garg Vipul -Principal author,
Glenys Poon-review of lit and write up,
Aprine Tan – data collection and patient managment,
Kein Boon Poon -consultant involved in management of patient, main guidance for write up.

Registration of research studies

Not required as no added intervention done for purpose of study.

Guarantor

Kein Boon Poon.

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