

Effort thrombosis of the subclavian-axillary vein

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ABSTRACT

Objective: The aim of this study is to outline the clinical patterns, diagnosis and the outcome of patients with 'effort' subclavian-axillary vein thrombosis.

Methods: The medical records of 7 patients diagnosed with 'effort' subclavian-axillary vein thrombosis between 1992 and 2000 at the Princess Basma Teaching Hospital, Irbid, Jordan were reviewed. Patients with secondary subclavian-axillary vein thrombosis (catheter related or secondary to thoracic outlet compression) were excluded.

Results: The clinical presentations of this condition

were swelling, pain, and cyanosis of the upper limbs. The presence of dilated superficial vein is a late sign. All patients were treated by non-fractionated heparin continued with warfarin with a favorable outcome.

Conclusion: Effort thrombosis of subclavian-axillary vein (Paget-Von Schroetter syndrome) is less reported in the literature contrary to secondary subclavian-axillary vein thrombosis. Early and effective anticoagulation constitutes the base of curative treatment. Prevention of recurrence is mandatory.

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Effort thrombosis of the subclavian-axillary vein, also called Paget-Von Schroetter syndrome is a rare condition, accounting for 1-2% of all cases of venous thrombosis.¹ It is called effort thrombosis, as it often follows excessive or unusual physical exercise. It is less known, as the advent of percutaneous technique brought regarding a large increase in the number of catheter-related thrombosis with a relative change in incidence between effort thrombosis and catheter-associated subclavian-axillary vein thrombosis.² The purpose of this study was to outline the clinical pattern and diagnostic features as well as the clinical course and outcome of this disease.

Methods. The clinical notes and radiological investigations of all patients admitted with the diagnosis of effort subclavian-axillary vein thrombosis to Princess Basma Teaching Hospital,

Irbid, Jordan were retrospectively reviewed. Seven cases of effort subclavian-axillary vein thrombosis were documented between January 1992 and December 2000, which represents 6% of all deep venous thrombosis of upper limbs found in the same period. All cases studied in regard to the clinical criteria shown in **Table 1**, in addition to investigations such as duplex ultrasound (US), venogram when necessary and hemostatic tests. Patients were reviewed at 6-months and one-year period of follow up with monthly clinical assessment and duplex US evaluation. Patients with secondary subclavian-axillary vein thrombosis (catheter related or secondary to thoracic outlet compression) were excluded.

Results. Clinical characteristics of the patients are summarized in **Table 1**. They were similar to

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other types of deep venous thrombosis of upper limb with essentially swelling, erythema, acute pain and sometime cyanosis. Most of our patients (6/7) presented late as evident by the presence of mild superficial venous circulation. Duplex US of upper limbs vessels confirmed the clinical diagnosis in all our 7 patients and demonstrated thrombosis of the subclavian vein in all patients with involvement of axillary vein in 6 patients. The investigation was completed by upper limb venography only in one patient due to doubt in diagnosis. Constitutional abnormalities of hemostasis (antithrombin III, protein C, proteins S, resistance to protein C, antiphospholipid antibodies and lupus anticoagulant factors) were studied in all patients and found to be within normal range. All patients were treated by intravenous non-fractionated heparin with therapeutic dose continued by oral anticoagulant (Warfarin) for 6 months. This was with contention by elastic bandages, active physiotherapy and professional rehabilitation, when needed. All patients were followed up by clinical examination and duplex US. Late evaluation at 6 months and one year of follow-up was favorable from clinical point of view, only 2 patients developed fatigue on exercise of the limb. Duplex US was normal in 5 patients, while in the remaining 2 patients who had fatigue on exercise of the limb, showed moderate stenosis of the subclavian-axillary vein without any new thrombus formation. Clinically, these 2 patients had good collateral venous circulation.

Discussion. Subclavian-axillary vein thrombosis is most commonly secondary to an indwelling catheter. However, it may also occur as a primary

event in young individuals ("effort thrombosis"). Its sudden clinical manifestations lead the patients to seek medical attention. Most patients with effort subclavian-axillary vein thrombosis are young with an average age of 30 years, with a 2:1 predilection of males over females.³ In our patients, the average age was 32 years, ranging from 22-42 years. The dominant extremity was reported to be involved in 60-80% of cases.^{4,5} In our patients, it was involved in 86% (6/7) of the cases. An antecedent event of strenuous or repetitive activity before the onset of symptoms was reported in 75% of patients.⁶ All our patients gave similar history. Effort subclavian-axillary vein thrombosis is the consequent of venous stasis with repetitive compressive trauma to the vein which may cause microscopic tears to the intimal layer.^{4,7,8} Aziz et al⁷ hypothesized that fibrosis and thickening of the vein wall and intimal proliferation developed after repetitive minor injuries to the vein with arm motion, such as 90° abduction, external rotation of scapulo-humeral articulation and alternate abduction-adduction of the upper limb. Compression of the subclavian vein in the costoclavicular space with hyper-abduction of the arm, hyper-extension of the neck, or downward and backward movement of the shoulder with subsequent narrowing of the vein has been proposed to explain the pathophysiology of this thrombosis.⁹ Some authors have suggested that the stress of exercise causes a temporary hypercoagulability.^{4,7,8} These mechanisms may be incriminated in 3 cases of our patients due to sustained physical effort with elevation and movement of abduction, adduction of the dominant upper arm with hyperextension of the neck. The slow progression of venous thrombosis allows time for the

Table 1 - Clinical findings of 7 patients with effort subclavian-axillary thrombosis.

Clinical criteria	Patient						
	1	2	3	4	5	6	7
Age	42	32	24	33	22	38	33
Sex	Female	Female	Male	Male	Male	Female	Male
Profession	Cleaner	Cleaner	Painter	Painter	Athlete	Cleaner	Plumber
Dominant arm	Right	Right	Right	Left	Left	Right	Right
Arm involved	Right	Right	Right	Left	Left	Right	Right
Edema	Positive	Positive	Positive	Positive	Positive	Positive	Positive
Pain	Positive	Positive	Positive	Positive	Positive	Positive	Positive
Cyanosis	Positive	Positive	Positive	Positive	Positive	Positive	Negative
Collateral circulation at presentation	Positive	Positive	Positive	Positive	Positive	Positive	Negative

development of collateral venous circulation. The hallmark of subclavian-axillary vein thrombosis is swelling of the involved extremity; this edema usually involves the entire arm and hand. In time, a variable percentage of patients will have obvious venous engorgement of the superficial collateral veins over the shoulder. The presence of superficial venous circulation reflects the late presentation of our patients. Most patients will eventually complain of pain in the affected extremity, described as "aching" or a feeling of tightness referred to the arm and axilla,¹⁰ and usually worsens with exertion. The diagnosis of effort subclavian-axillary vein thrombosis is based on the clinical presentation of upper extremity swelling, venous engorgement, and pain, relatively of sudden onset in a young physical worker with absence of other etiology being the origin of venous thrombosis. Diagnostic studies are indicated to confirm the diagnosis and to determine the extent of the thrombus. Duplex US is the diagnostic modality of choice with excellent specificity compared with venography. Koksoy et al¹¹ found that duplex scanning had a sensitivity of 94% and a specificity of 96% compared with venography. Advances in US technology (namely color flow scan) and adjunctive use of indirect criteria for proximal occlusion (distended, incompressible vein with poor augmentation of flow by compressive maneuvers) may improve sensitivity. A distance from the acute phase, the retrospective diagnosis is made by evidence of valvular incontinence and vein wall thickening. Duplex US allows the diagnosis and follow-up of the patients simply, non-invasively and repeatedly. Venography is still the "gold standard" in evaluating subclavian-axillary vein thrombosis, and it is required if doubt regarding diagnosis still persists or if surgery or endovascular interventions are contemplated.¹² The other diagnostic modalities, such as continuous wave doppler examination and impedance plethysmography have largely been supplanted by duplex US. Magnetic resonance angiography is specific for complete occlusion of the subclavian-axillary vein, but it has poor sensitivity, especially for non-occlusive thrombi and may also missed short segment occlusion.^{13,14} Radionuclide venography is useful in detecting the presence of thrombus, but does not precisely define the extent of the clot.¹⁴ Computed tomography has also been used to detect subclavian-axillary vein thrombosis, but unfortunately, comparison studies with venography are insufficient to determine its specificity and sensitivity. Due to its cost, relative accuracy, and ease of performances, venous duplex US remains the diagnostic tool of choice. Pulmonary embolism has been variously reported in patients with subclavian-axillary vein thrombosis, overall, the symptomatic pulmonary embolism rate is 2%.³ Hingorani et al³ found no difference in the rate of pulmonary

embolism between primary or secondary subclavian-axillary vein thrombosis, their overall incidence was 7% regardless of the underlying cause. None of our patients develop symptomatic pulmonary embolism. Post-thrombotic sequelae are the principal late complications of effort thrombosis. It may be symptomatic with pain or mild edema on exercise. This complication seems to be more in effort thrombosis than secondary thrombosis of subclavian-axillary vein.¹⁵ Optimal management of effort subclavian-axillary vein thrombosis is still surrounded with many controversies.¹⁶ Treatment options include anticoagulation therapy, catheter-directed thrombolysis, and surgical intervention. Anticoagulation is based upon heparin therapy with a therapeutic dose started early with the confirmation of the diagnosis and followed by oral anticoagulants for a period of 3-6 months. Although earlier reports suggested that thrombolytic therapy was insufficient in treating effort thrombosis,^{4,17} more recent reports indicated that there is a definite role for thrombolysis in selected cases especially acute thrombosis of less than one-month duration.¹⁸⁻²⁰ Surgical treatment by removing the intravascular clot, revising the anatomy of the costoclavicular space or resection the first rib should be reserved for patients in whom there is specific indication such as of thoracic outlet syndrome.^{5,6,20}

In conclusion, effort thromboses are rare. Multiple mechanisms mainly repeated physical exercise underlies their pathophysiology. Contrary to secondary thrombosis, they are rarely embolic but late sequelae are more common than secondary thrombosis, even in the presence of important collateral circulation. Early and effective anticoagulation constitute the base of curative treatment. Prevention of recurrence and late sequel is achieved by elastic contention, active physiotherapy and professional rehabilitation, in patients at risk.

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