Presentation

A 24 YO male, a ping-pong player, presented to the ER with acute onset of right upper extremity pain (his dominant arm), following a strenuous activity the day before.

Background

- **Tetraplegia** followed a car accident in 2007
- **Asthma** since childhood, was discharged from our department 4 days ago after a prolonged hospitalization with URTI & Asthma exacerbation.
- **Pressure ulcer** at sacrum – grade I-II
Vital signs

BP 115/40   HR 82 BPM, regular   SAT 96% RA   TEMP 37.6°C

Physical examination findings

- Lung, Heart, Abdomen: normal findings
- Extremities: Right arm & armpit edema with slight cyanosis of fingers.

ECG

NSR, regular, normal axis, no signs for acute ischemia
Diagnostic studies

Laboratory

- CBC – Hb 14.5 g/dL (13.5-17.5), **WBC 16.1** (4-11), Neut 79% (40-75)
- Coagulation – INR 1.05 (0.85-1.2), PLT 244 (150-400)
- Chemistry – CRP 6.7 mg/L (-6); liver & renal intact; electrolytes are normal
- **D-dimer 1254 ng/mL** (-500) – In the previous hospitalization it was 450 ng/mL.

Imaging

**US-D** – Thrombosis of the Rt axillary & subclavian veins, normal flow at the left arm.
**CTA** chest – Pulmonary emboli at the 3\(^{rd}\)/4\(^{th}\) branches of the Rt lung.
**CT-V** upper extremities – Thrombosis of the Rt brachial, axillary & subclavian veins.
Venography

Notice the amount of collaterals
DVT of RT upper limb - DD

- central venous cannulation (d/t Central-line catheter)
- Hypercoagulability state
- Trauma
- Malignancy
- Paget-Schroetter syndrome / effort-induced thrombosis
Paget-Schroetter syndrome

Thrombosis of upper extremity deep veins due to anatomic abnormalities of the thoracic outlet
Also called “Primary Effort Thrombosis”

Epidemiology

- 1-4% of all cases of Upper extremity DVT
- M:F ratio is 2:1
- Mean age- early 30s
- Usually the dominant upper extremity
- Strenuous upper extremity activity-60-80%
- Anatomic abnormalities of the thoracic outlet (congenital, acquired)
Paget-Schroetter syndrome
Paget-Schroetter syndrome - Pathogenesis

- Thoracic outlet anomaly
- **BUT!!!**
- Anatomic abnormality is not necessary to produce injury to the vein.
- Repetitive injury causes inflammation and fibrosis, which eventually leads to thrombosis.
- Collateral pathways - important for diagnosis
Treatment

- **Anticoagulation** - Low molecular weight heparin started at his first day 1mg/kg * 2/d

- **Chatheter-directed Thrombolysis** – for 48 hrs. no success in opening the veins but increased flow in collaterals.

- **Thoracic outlet decompression (surgical)** wasn’t done.

- **Heparin induced thrombocytopenia (HIT)** - PLT drop almost by 50% (267->144) after 7 days of treatment -> ELISA against heparin-PF4 complexes was positive.

- **LMWH switched to fondaparinux and later on Warfarin was initiated.**
Follow up (Hematology)

- Anti Beta-2 Glycoprotein 1 (AB2G1) IgG – 62.2 U/ml (-15)
- dRVVT, AB2G1 IgM, anti Cardiolipin IgM & IgG all negative
- Secondary AB2G1 IgG after 5 months – 24 U/ml (-15)
- US-D – after few months, no thrombosis in the right upper limb
- Continues daily Warfarin (7.5 mg – 10 mg) in therapeutic INR (2-3)
- post-thrombotic syndrome at 5 year (53% Vs 12-25%)

Antiphospholipid syndrome (APLS)?
That was only 7 years ago...
Back to his life...a year after his DVT

May 2014 – 5th time Israeli champion

March 2014
Men’s teams in class 1-2
Bronze medal
Summary & key points

DVT of upper extremities – a rare disorder

Paget-Schroetter as an epiphenomena or is it APLS ???

Anti Beta-2 Glycoprotein 1

Treatment option – invasive Vs non-invasive

How further should the treatment continues?
A CASE REPORT: A YOUNG WAITER WITH PAGET-SCHROETTER SYNDROME

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Over Rehearsed: A Cellist With Paget-Schroëtter
Bryan P. Yan, Thomas J. Kieman, Vishal Gupta, Robert M. Schainfeld and Joseph M. Garasic

Circulation. 2008;118:e160-e161

Thank You