**Case Vignette**

A 13-year-old male student, who is an only child, from Manila was brought in for consult at the emergency department due to sudden onset of severe left flank pain characterized as piercing associated with numbness and weakness of the left thigh and leg leading to difficulty in ambulation. He has no fever, cough, difficulty of breathing, nausea, vomiting, changes in appetite, changes in sensorium, bowel movement changes or urinary changes. He has history of easy bruisability since birth and has a history of prolonged bleeding after a dental extraction and after being stabbed by a pencil at school in the arm. Also, he had recurrent episodes of right knee pain. His maternal grandfather died of an unrecalled bleeding disorder. He denies smoking, taking any medications, alcoholic beverage drinking and use of illicit drug use.

Initial PE on consult:

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| --- | --- |
| General appearance | Awake, in pain |
| Vital Signs | BP 120/80, HR 100, RR 24, Temp: 36.5oC |
| HEENT | Anicteric sclerae, pink palpebral conjunctivae, no palpable lymphadenopathies. |
| Chest/Lungs | Symmetrical chest expansion, clear breath sounds |
| Cardiovascular | Adynamic precordium, regular rate and rhythm, distinct S1 and S2, no murmurs, PMI at 5th ICS left midclavicular line |
| Gastrointestinal | Flat abdomen, normoactive bowel sounds, tympanitic on percussion, (+) tenderness on left flank. (+) aggravation of left flank pain on extension of left lower extremity. no palpable masses. |
| Extremities | Full and equal pulses. No bipedal edema. Prefers to keep left lower extremity in flexed position at the hip joint.  (+) swollen, warm to touch, slight tenderness on right knee |
| Neurologic Exam | (+) 50% sensation on the anteromedial aspect of the left thigh and leg.  (+) able to dorsiflex and plantar flex left foot but unable to flex hip against resistance and unable to bend knee. however the motor examination on the left lower extremity is limited due to pain  (+) absent patellar reflex on the left |

**Initial Labs done on consult**:

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| **Hgb** | 146 g/L |
| **Hct** | 44% |
| **RBC** | 5.03 x1012/L |
| **MCV** | 88.2 fL |
| **MCH** | 29.0 pg |
| **RDW** | 14.4% |
| **WBC** | 9 x109/L |
| **Neutrophil** | 74% |
| **Lymphocyte** | 12% |
| **Monocyte** | 9% |
| **Eosinophil** | 4% |
| **Basophil** | 1% |
| **Platelet** | 383 x109/L |

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| --- | --- |
| **PT reference** | 12.5 secs |
| **PT** | 13.2 |
| **INR** | 1.05 secs |
| **PTT reference** | 28.2 secs |
| **PTT** | 80.8 secs |

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| Crea | 49 umol/L | Na | 135 mmol/L |
| AST | 35 U/L | **K** | 3.9 mmol/L |
| ALT | 29 U/L | **Cl** | 107 mmol/L |
| Alb | 3.8 g/dL | **Ca** | 4.2 mg/dL |
| TB | 1.5 mg/dL | **Mg** | 0.81 mmol/L |
| DB | 0.5 mg/dL | **LDH** | 100 U/L |
| IB | 1.0 mg/dL |

**Subsequent Labs done:**

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| **PTT reference** | 28.2 secs |
| **PTT** | 80.8 secs |
| **PTT with mixing** | 27.0 secs |
| **1 hour incubation** | 27.0 secs |
| **2 hour incubation** | 27.3 secs |

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| **Fibrinogen** | 250 mg/dL |
| **Factor 8 assay** | 2% |
| **Factor 9 assay** | 80% |

Whole abdominal ultrasound: (+) iliopsoas bleed, left; Normal ultrasound of the Liver, Spleen, Pancreas, Abdominal Aorta and Para-aortic areas and Kidneys