DIAGNOSTIC CHALLENGES IN A CASE LIFE-THREATENING THROMBOSIS AND FEVER OF UNKNOWN ORIGIN

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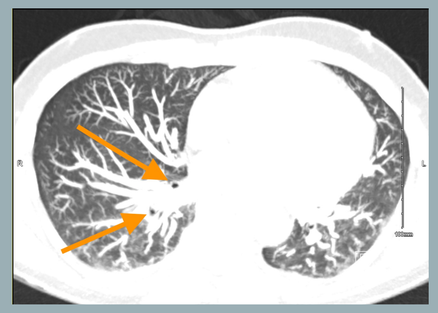
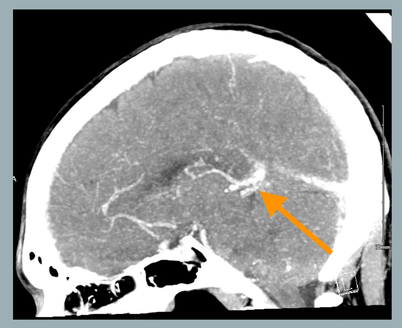
Introduction

Inflammatory diseases can often be difficult to diagnose and rely upon the exclusion of other potential pathologies. A delay in diagnosis can result in greater morbidity for the patient. We present the diagnostic challenges in the case of Behcet's disease who presented with recurrent venous thromboembolism (VTE) and fevers of unknown origin (FUO).



CT pulmonary angiogram demonstrating segmental and subsegmental pulmonary emboli of the right lower lobe (orange arrows)



CT venogram demonstrating nonocclusive central venous sinus thrombosis (orange arrow)

Ferritin	709 µmol/L (23-393)
Anti Xa level (taken as a through while on 1mg/kg of enoxaparin)	0.5 IU.ml
Beta-2-glycoprotein	5.1 U/ml (0-6.99 U/ml)
IgG cardiolipin antibodies	Low positive
Dilute Russell viper venom test	1.41 (0-1.26)
Paroxysmal nocturnal hemoglobinuria screen	Negative
JAK2 V617F mutation	Not detected
Anti thrombin activity	1.01
Anti dsDNA (ELISA)	34 (0-9.99)
Anti dsDNA (Crithidia assay)	Negative
Anti centromere/La/RNP/Smith/Jo/Scl	Negative
IL6	7.29 pg/ml (0.09-7.26)
Anti-GBM	Negative
ANCA	Negative
IgG	12.3 g/L (6.26-14.9)
IgA	4.76 g/L (0.62-2.90)
IgM	0.85 (0.47-1.82)
QuantiFERON assay	Negative
Blood cultures x7	Negative
HIV Ab/Ag assay	Negative
HepBsAb	Negative
Hep C antibody	Negative
Schistosomiasis (ELSIA)	Negative
TPMT result	34 (normal)
Malaria Rapid diagnostic test	Negative
Rickettsia serology	Negative
Syphilis serology	Negative

Case Description

A of a 53-year-old gentleman of Irish Caucasian ethnicity presented with a history of fevers and recurrent VTE at a university hospital in Dublin, Ireland. Past medical history includes schistosomiasis which was treated following a trip to sub-Saharan Africa. Our patient was previously diagnosed with a provoked deep vein thrombosis (DVT). He went on to experience 4 subsequent episodes of VTE, including DVT, pulmonary embolism (PE), and cerebral venous sinus thrombosis (CVST) while on different forms of anticoagulation. On each of these occasions, there was a concern for sepsis due to fevers >38 degrees and a CRP >200.

The infectious workup included routine labs, blood and urine cultures, CT abdomen/pelvis, echocardiogram, and PET CT, all of which were unrevealing.

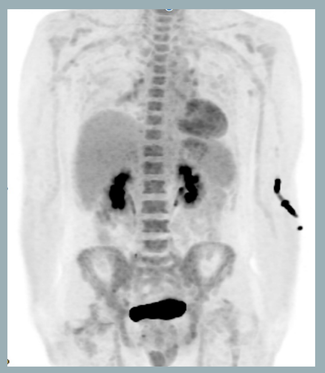
However, a focused clinical exam revealed evidence of subtle scrotal and oral ulceration, pustulation, and erythema at several sites in his upper limb following venesection and cannulation. In this context, a diagnosis of Behcet's disease was considered.

Discussion

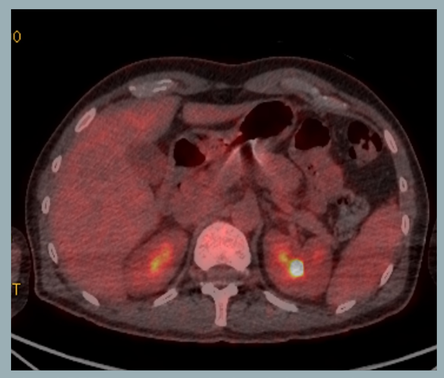
A diagnosis of Behcet's can only be confidently made after the exclusion of other potential etiologies. In this case, we had to consider a broad range of infectious (malaria, schistosomiasis, rickettsial disease, endocarditis) and noninfectious diseases (malignancy, anti-phospholipid syndrome, myeloproliferative disorders, paroxysmal nocturnal hemoglobinuria).

A delay in diagnosis comes at the cost of increased morbidity and mortality for the patient. A detailed history and clinical exam are key, in addition to a high index of suspicion.

Following induction of high-dose steroid, our patient is doing very well on maintenance Adalimumab. From an anticoagulation perspective, he is warfarinised and has not had any further episodes of VTE.



Coronal PET scan deomstrating physiological uptake of tracer



Axial PET scan through the abdomen demonstrating physiological uptake with some concern for renal infarction or infection