Moroccan clinical case

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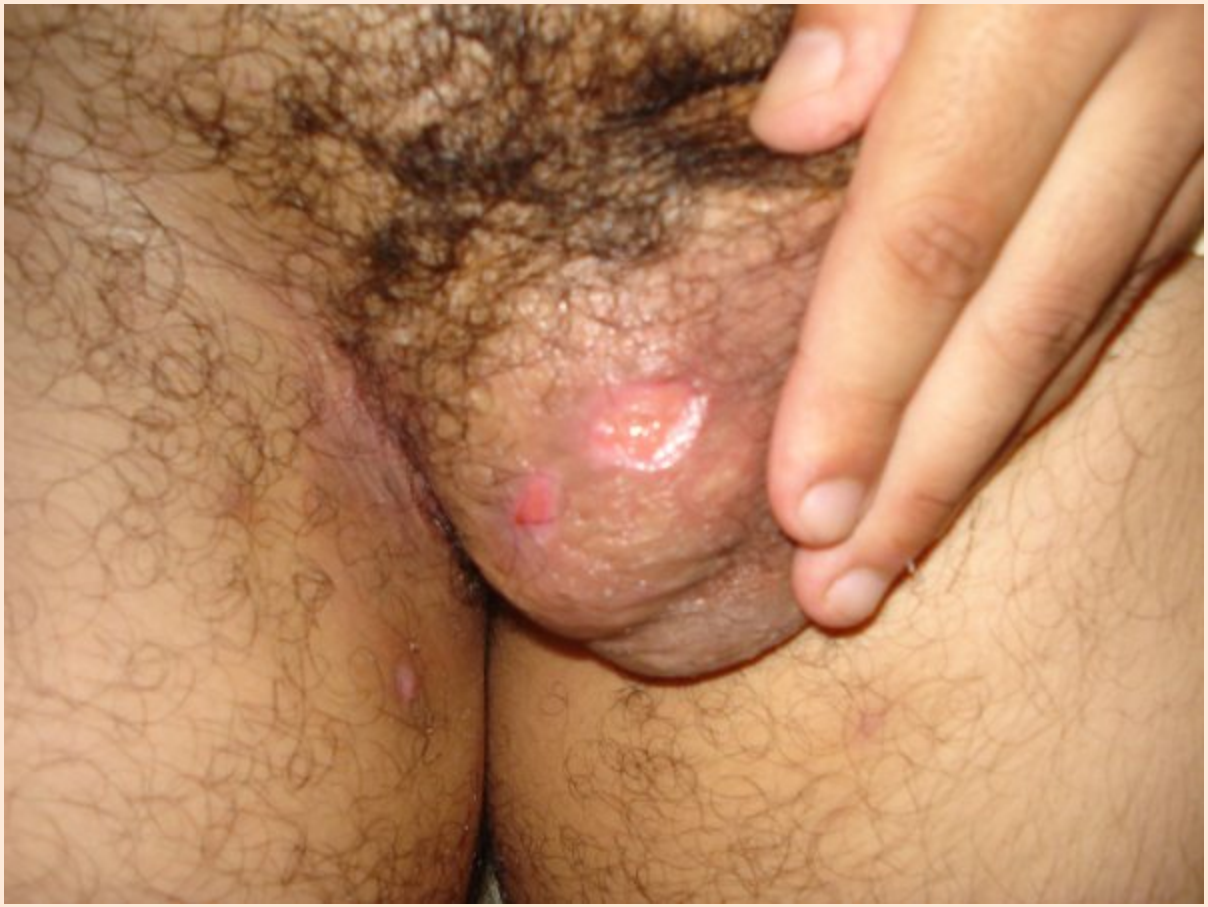
- A 40-year-old Moroccan man
- Three days ago: redness, warmth, swelling, pain in the left leg
- No others functional symptoms

Physical examination

- GCS : 15
- Temperature: 37°6 C
- Blood pressure 130/70 mmHg
- pulse rate 90 beats/mn



- Oral and genital aphtosis
- Pseudofolliculitis
- No other abnormalities

(no symptoms of pulmonary embolism)

Paraclinical investigations

- Vein doppler of the left leg : deep vein thrombosis arriving until the primitive iliac vein
- Electrocardiography – chest radiography : normal
- Blood numeration – CRP – sedimentation rate – hepatic parameters - creatinine : normal
- Abdominal Echography : normal

What is your diagnosis ?

- Lupus ?
- Inflammatory bowel disease ?
- Behçet disease ?

➔ antinuclear antibodies negative
➔ no transit abnormalities

Behçet’s disease

- Criteria :
  bipolar aphtosis
  pseudofolliculitis

- Other classical finding :
  deep vein thrombosis

What is the management ?

- Corticosteroids
- Immunosuppressive therapy (azathioprine or cyclophosphamide)
- ± anticoagulation therapy

Eular recommandations for the treatment of vascular behçet disease

Can we consider that diagnosis and therapeutical mangement of our patient were good and complete?

Etiologic diagnosis of deep vein thrombosis

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some paraclinical investigations must be performed systematically

The most important explorations in deep vein thrombosis

- Blood numeration
- Sedimentation rate
- C reactive protein
- Chest radiography and abdominal ultrasound
- Serum Proteins electrophoresis
- Proteinuria

In our case

- Blood numeration : normal
- Sedimentation rate and CRP normal
- **Serum protein : 50 g/l**
- **Serum Albumin : 20 g/l**
- **Proteinuria : 11 g/day**

➔ nephrotic syndrome

Causes of nephrotic syndrome ?

- No diabetes
- No signs of lupus
- No signs of amylodosis
- **Kidney biopsy :**

  extramembranous glomerulonephritis

Is the extramembranous nephritis due to Behçet disease?

Causes of secondary extramembranous nephritis

- infections : hepatitis B, syphilis, malariae
- systemic diseases : lupus, RA, sarcoidosis
- drug intake : D penicillamine, non steroidien anti-inflammatory, captopril…
- neoplastic : lung, breast, lymphoma…

The final diagnosis

- Deep vein thrombosis with 2 major causes :

  Behçet disease

  Nephrotic syndrome (extramembranous nephritis)

Treatment ?

- Deep vein thrombosis : Anticoagulation +++
- Nephrotic syndrome : ACE inhibitors
  Furosemide
  Statins
- Corticosteroids
- Immunosuppressive therapy : which one?

- Deep vein thrombosis in Behçet disease :

  azathioprine or cyclophosphamide

- Extramembranous nephritis :

PONTICELLI’s protocol (chlorambucil – corticosteroids)

In our case :

Protocol ponticelli

6 months later

- Complete normalisation of clinical and paraclinical findings :

➔ serum protein and albumin

➔ negative proteinuria

➔ no aphtosis

The most important messages

- Deep vein thrombosis may have many associated causes at the same time
- the etiologic exploration must be complete even if the diagnosis appears to be clear at the initial presentation
- Glomerulopathy is rare but may exist in Behçet disease (amylosis – proliferative nephritis +++)

Thank you for your attention