Behçet's disease

This is a vasculitis of unknown aetiology that characteristically targets small arteries and venules.

There is a strong association with HLA-B51.

Behçet's disease was thought to predominantly affect males, but current epidemiologic data show a more equal male to-female ratio. Males tend to have a more severe clinical course, with increased disease associated morbidity

Clinical features:

Oral ulcers are universal. Unlike aphthous ulcers, they are usually deep and multiple, and last for 10–30 days, occur more than 3 times/year.

Genital ulcers are also a common problem. It heals with scar.

The usual skin lesions are erythema nodosum or acneiform lesions.

Ocular involvement is common and may include anterior or posterior uveitis or retinal vasculitis.

Neurological involvement: mainly involves the brainstem.

Vascular involvement either arterial or venous, superficial or deep, thrombosis or aneurysm.

Criteria for diagnosis 2010

Oral aphthosis	2
Genital aphthosis	2
Ocular manifestations	2
Skin manifestations	1
Vascular manifestations	1
Central nervous system involvement	1
Positive pathergy test	1

Need at least 4 points for diagnosis.

The diagnosis is primarily made on clinical grounds.

Pathergy test, which involves pricking the skin with a needle and looking for evidence of pustule development within 48 hours.

Treatment:

Oral ulcer and genital ulcer: colchicine

Mucocutaneous: Low-dose oral methotrexate (2.5 to 25 mg/wk) and low dose prednisone

Arthritis: NSAID, colchicine, prednisolone, MTX

Ocular: topical or systemic steroid, immunosuppressant

GIT: steroid, sulphasalazine, azathioprine

CNS and vascular: steroid and immunosuppressant

In patients with BS, deep vein thrombosis is thought to result from inflammation of the vessel wall rather than hypercoagulability. So the use of anticoagulant in vascular thrombosis is still controversial.

Septic arthritis:

Septic arthritis is the most rapid and destructive joint disease.

Pathogenesis

Septic arthritis usually occurs as a result of haematogenous spread from infections of the skin or upper respiratory tract; or from direct puncture wounds.

In adults, the most likely organism is Staphylococcus aureus, particularly in patients with RA and diabetes.

Staphylococcus epidermidis is common in prosthetic joint.

In young, sexually active adults, gonococcus may be responsible.

Gram-negative bacilli is important cause among the elderly and intravenous drug users.

Risk factors

Include

1- increasing age,

2- Pre-existing joint disease (principally RA).
3- Diabetes mellitus.
4- Immunosuppression (by drugs or disease)
5- Intravenous drug misuse.

Clinical features

The usual presentation is with acute or subacute monoarthritis and fever. The joint is usually swollen, hot and red, with pain at rest and on movement

Patients with pre-existing arthritis may present with multiple joint involvement.

Disseminated gonococcal infection occurs in up to 3% of patients with untreated gonorrhoea. This usually presents with migratory arthralgia, low-grade fever and tenosynovitis, which may precede the development of an oligo- or monoarthritis. Painful pustular skin lesions may also be present.

Investigations

The pivotal investigation is joint aspiration but blood cultures should also be taken. The synovial fluid is usually turbid or bloodstained but may appear normal.

Synovial fluid should be sent for Gram stain and culture; cultures are positive in around 90% of cases but the Gram stain is positive in only 50%. In contrast, synovial fluid culture is positive in only 30% of gonococcal infections, making it important to obtain concurrent cultures from the genital tract.

There is a leucocytosis with raised ESR and CRP in most patients, but these features may be absent in elderly or immunocompromised patients.

Serial measurements of CRP and ESR are useful in following the response to treatment.

Management:

The patient should be admitted to hospital for pain relief and administration of parenteral antibiotics.

Flucloxacillin (2 g IV 4 times daily) is the antibiotic of first choice pending the results of cultures.

If there is reason to suspect meticillin-resistant Staphylococcus aureus (such as a known carrier), vancomycin should be used instead while awaiting cultures.

If a Gram-negative infection is suspected, gentamicin or vancomycin should be considered as first-line treatments. Cephalosporins are a potential alternative for Gram-negative infections

It is traditional to continue intravenous antibiotics for 2 weeks and to follow this with oral treatment for another 4 weeks.

Joint aspiration should be performed using a large-bore needle once or twice daily. If this is not possible, arthroscopic or open surgical drainage may be needed performed.

Regular passive movement should be undertaken from the outset, and active movements encouraged once the condition has stabilized.

Read more in:

1- Davidson's Principles and Practice of Medicine, 23rd edition
2- Kelley & Firestein's Textbook of Rheumatology, 10th edition