DEFINITION

1. Behçet's syndrome is a chronic, often progressive, disease, characterised by recurrent aphthous stomatitis of the mouth associated with genital ulceration and eye disease (especially retinal vasculitis).

CLINICAL MANIFESTATIONS

2. The full syndrome comprises oral and genital ulceration, ocular lesions and a characteristic pyoderma. In many cases the clinical picture is ultimately dominated by changes in the central nervous system or, less commonly, by pulmonary or cardiac involvement.

3. The first manifestation is usually oral or genital ulceration followed after an interval of days or months by iritis and then by involvement of other organs.

4. The mouth ulcers are rounded or oval and are sharply demarcated with a yellowish floor and a bright red areola. They may increase in size for weeks and heal only after several months. They may occur anywhere in the mouth and may extend to the oesophagus. Pain and tenderness may be severe and dysphagia may be a problem.

5. The genital ulcers occur mainly on the scrotum, around the root of the penis or on the labia majora.

6. Skin lesions are present in the majority of cases at some time. The rash is of a follicular and non-follicular pustular type on the trunk and limbs but tends to favour the genital area.

7. The ocular features are photophobia, conjunctivitis and uveitis, often with loss of vision from vitreous opacities. Sometimes posterior uveitis and optic neuritis may occur.

8. Other systems may be involved leading to arthralgia, thrombophlebitis, brain stem syndrome, organic confusional state, manifestations of meningoencephalitis, and rare manifestations of pulmonary or cardiac involvement.

AETIOLOGY

9. This is uncertain. There is a genetic element, with a higher incidence in some geographical areas, some familial cases and associations with HLA types, particularly HLA-B5.

CONCLUSION

10. Behçet's syndrome is a combination of ulceration of the mouth and genitalia together with iritis and skin lesions. Lesions in other systems may also occur. The aetiology is unknown but genetic factors are important.

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