SELF - ASSESSMENT PROGRAMME

A 27 year old male university student complained, for one week, of painful ulcerations in the mouth, redness of eyes, high grade fever and pain in both the calves. A day later he also developed erythematous painful nodules on the dorsa of both feet and later, on both the upper and lower extremities. Pain with swelling of both knee and ankle joints was also noticed for 2 days. Treatment with antibiotics and analgesics/antipyretics did not affect the course of the disease.

The patient, a well built young man, had multiple, discrete and confluent, tender, oval and circular shallow ulcers on the tonsils, uvula, soft palate and oropharynx; the ulcers having a white necrotic base and an erythematous halo.

There was bilateral conjunctivitis and tender swelling of both knee and ankle joints. Calf tenderness was present.

Multiple warm, erythematous, tender nodules, 1½–2 cm diameter were present on both the feet, legs and thighs. Similar but fewer lesions were present on both the arms.

1. What is the most probable diagnosis ?
   a) Acute follicular tonsillitis with erythema nodosum
   b) Diphtheria with erythema nodosum
   c) Behcet's syndrome
   d) Secondary syphilis
   e) Agranulocytic ulcers with drug induced erythema nodosum.

2. What further clinical signs would you specifically look for ?
   a) Genital lesions
   b) Generalised lymphadenopathy
   c) Central nervous system involvement
   d) Detailed ophthalmic examination
   e) Evidence of thrombophelebitis

On further examination a round, crusted ulcer, 1 cm in diameter with an indurated tender base, was found on the scrotum and a history of recurrent scrotal ulceration over the last 7 years was obtained. Palpation below the popliteal fossae revealed a tender, firm, cord like structure on each side. There

was no generalised lymphadenopathy or CNS involvement; slit lamp examinations revealed bilateral active nongranulomatous iridocyclitis.

3. What investigations would be diagnostic ?

   a) V. D. R. L.

   b) Throat swab for culture and sensitivity

   c) ASLO Titre

   d) Intradermal saline test for skin hyper reactivity

   e) Skin biopsy

   f) Total & Differential leuckocyte counts

VDRL was non reactive. Throat swab culture revealed normal flora. ASLO titre was 166–250 units and there was no pustule formation on intradermal injection of saline. Skin biopsy from a nodular lesion showed hypersensitivity vasculitis. The patient had a polymorphonuclear leucocytosis.

A diagnosis of Behcet's syndrome was established.

4. What further investigations would you like to do ?

   a) Serum C9, CRP, Immunoglobulin profile

   b) E. C. G.

   c) Chest X-ray

   d) E. E. G.

   e) Blood fibrinogen level & euglobulin lysis time

5. How would you treat the patient ?

   a) Systemic corticosteroids

   b) Local tetracycline and/or corticosteroids

   c) Oral colchicine

   d) Levamisole.

6. What is the likely prognosis in this patient ?

   a) Unpredictable with relapsing course

   b) Good

   c) Poor

   d) Fatal

ANSWER

1–c, Painful red nodules with fever and arthritis may occur in erythema nodosum but the shallow buccal ulcerations

do not fit in with an acute follicular tonsillitis or diphtheria. The total clinical picture is not suggestive of secondary syphilis. The rare possibility of agranulocytic ulcers with coexistent drug induced erythema nodosum cannot be entirely ruled out from the data, though it seems unlikely. Behcet's syndrome is thus perhaps the most probable diagnosis with characteristic oral ulcerations, eye involvement and erythema nodosum-like skin lesions. Thrombophlebitis and arthritis seen in this patient are also commonly present in this disease complex.

2–a,c,d,e, With a diagnosis of Behcet's syndrome in mind one would look for involvement of CNS, eyes and thrombophilebitis.

3–d, Skin hyperreactivity in Behcet's syndrome can be demonstrated by intra dermal injection of normal saline producing sterile pustules at the injection site.

Total leucocyte count would help in excluding agranulocytosis and a skin biopsy would clinch a diagnosis of erythema nodosum.

4. All the above would be useful. Serum C9 CRP and IgA are raised in many active cases of Behcet's syndrome. ECG and chest x-ray would respectively help to determine cardiac or pulmonary involvement. EEG may sometimes show abnormalities in patients found normal on clinical neurological evaluation. In cases where active thrombophlebitis is present blood fibrinogen levels are raised and euglobulin lysis time is elevated.

5. In this case, due to iridocyclitis, systemic steroids and levamisole were given though other modes of treatment are not without merit.

6. The disease is characterised by exacerbations and remissions of unpredictable duration.

Comments

Behcet's syndrome is a rather uncommon triad of oral and genital ulcers and ocular involvement. Not uncommonly there are other associated features like pustules and erythema nodosum lesions, thrombophlebitis, arthralgia/arthritis and central nervous system involvement. Rarely cardiac, pulmonary and gastro-intestinal complications may be present. Curth suggested that 2 of the following criteria viz, oral ulcers, genital ulcers, eye involvement, must be fulfilled before a diagnosis of Behcet's syndrome can be accepted.

The course of the disease is variable - often progressive with relapses and remissions and may even finally ending fatally. The fatality is often related to oculo-neurological involvement

rather than muco-cutaneous syndrome. Laboratory affords limited help in confirmation of diagnosis, though a useful bed side procedure is the development of a pustule at the site of an intradermal injection or prick.

Behcet's syndrome is a difficult therapeutic problem with unpredictable outcome. Topical and systemic corticosteroids and/or antibiotics have been used with inconsistent results. Levamisole (levo-tetramisole) has also been employed with variable results.

References

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