BEHECETS' SYNDROME
(A case report)

K. C. VERMA AND KRISHANBIR SINGH

Behecets' syndrome is a rare disorder of unknown etiology. It consists of recurrent ulcerations of oral and genital mucosa and relapsing iritis. It is a potentially dangerous disease and runs a progressive and chronic course. Since its original description by Behecet in 1937, a number of case reports have appeared in the literature from all parts of the world. A case of Behecets' syndrome with a brief review of literature and clinical features is presented.

Case history

A, 55 years, male farmer was admitted in the Skin Ward of Medical College and Hospital, Rohtak in February, 1970. His complaints at the time of admission were recurrent ulcers in the mouth and on the skin of scrotum and penis for the last 5 years. Since the same duration he had been having transitory attacks of nodular eruptions on the extensor aspects of fore-arms and thighs. With each such attack he had mild to moderate degree of fever and joint pains. On occasions he developed pustular lesions on the skin particularly over the traumatic or injection sites.

After about 3 years of onset of the disease the patient developed gradual loss of vision first from the left eye and then from the right. He denied any history of intake of drugs previous to the disease or any history of syphilis. There was no positive family history.

Department of Skin & V.D.,
Medical College and Hospital, Rohtak
Received for Publication on 23—2—1972

Examination

On examination the patient showed multiple superficial ulcers, 1–2 cms in size all over the mucous membrane of the mouth. The genital ulcers were slightly bigger and punched out in appearance (Fig. No. 1). Multiple scars were present over the skin of the scrotum and penis. Patient had a few small tender nodules on the back of the elbows and fore-arms on both sides. Small pustules were seen at injection sites (Fig. No.2).

Examination of the eyes revealed, nebular opacities in cornea of both sides, fine pigmentation at lower half of both the corneae and optic atrophy more marked on the right than the left side. Vision was 6/60 in the right eye and 6/18 in the left eye. Field of vision was reduced on both sides.

General physical examination did not reveal anything significant except moderate anemia. Nervous system, C.V.S. Respiratory system and Abdomen were all normal.

Investigations

Hb. 10g%, T.L.C. 8300/cmm. D.L.C. P60%, L36%, M2%, E2%.

E.S.R. 85 mm. Platelet count 350000.

Urine and stools–N.A.D.

Screening chest–N.A.D.

X-ray chest : Showed a calcified shadow in the upper zone of the left lung.

Fig. 1

Shows multiple, deep, punched out ulcers on the skin of scrotum.

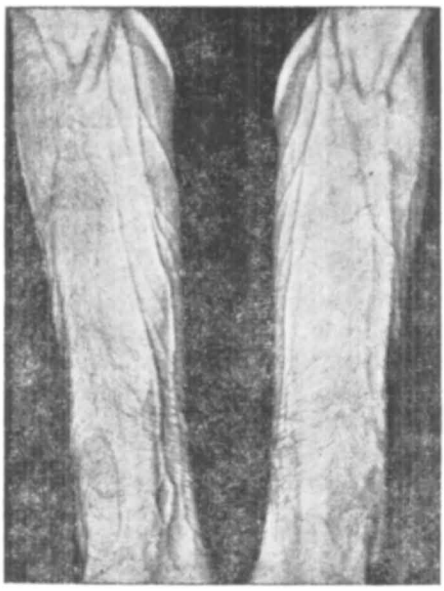

Fig. 2

Shows pustules at the sites of intravenous injections on the left fore-arm.

P.B.F. : Showed hypochromic microcytic anemia.

L.E. cell phenomenon was negative.
E.C.G.–Normal.
C.S.F.–Normal.

Biopsy from scrotal ulcer was non-specific and showed patchy acanthosis and heavy infiltration of upper dermis with lymphocytes and neutrophils particularly around the vessels.

Discussion

The association of recurrent oral and genital ulcers with iritis and hypopion was first described by Planner and Romanovsky in 1922 and Admantiades in 1931. In 1937 Behecet coined the name of this disease. Originally the syndrome consisted of the triad of oral and genital ulcers with recurrent iritis. Later various other clinical findings were added to this condition. Thrombophlebitis was described by France et al. in 1961. Neurological manifestations by Pallis and Fudge in 1956; Polyarthritis by Stranchan and Wigzell in 1963 and Cardiac manifestation like pericarditis by P. D. Lewis in 1954. A characteristic pustulation at traumatic sites was described by Rook et al.[12] Cases from India were reported by Misra in 1963 Jain in 1962 and Nanda in 1964 and many other workers. In our case oral and genital ulcers, corneal opacities, optic atrophy thrombophlebitis and characteristic pustulations at injection sites were present.

Etiology of the condition is still unsolved. Theory of focal sepsis and chronic respiratory infection was put forward by Thomas in 1947. The presence of circulating antibodies against the buccal mucosa make autoimmunity a possibility (Lehner in 1967). Sezer in 1953 isolated a virus from the blood and urine of the patient of Behecets' disease. Sensitization to sulfa drugs has been blamed as a cause of the disease by Jain in 1962.

Summary

A case of Behecet syndrome is presented. Literature and clinical features are briefly reviewed.

Acknowledgment

We are very thankful to Dr. Chander Prakash, Professor of Medicine and Medical Superintendent of Medical College and Hospital for permitting us to publish this case report.

REFERENCES

1. Adamantiades B: Ann Occulist (Paris) 168 : 271, 1931. Cited by 3.
2. Behecet H: Derm Wschr 105 : 1152, 1937. Cited by 6.
3. Fowler TJ: Behecet's Syndrome with neurological manifestations, Brit Med J 2 : 473, 1968.
4. France J: Medicine (Baltimore) 30 : 335, 1951. Cited by 7.
5. Jain ACS: Behecet Syndrome, J Ind Med Assoc 38 : 555, 1962.
6. Lehner T: Behecet Syndrome and autoinmunity Brit Med J 1 : 465, 1967.
7. Lewis PD: Behecet disease and carditis Brit Med J 1 : 465, 1967.
8. Misra NP: Behecets Syndrome, J Ind Med Assoc 40 : 469, 1963.
9. Nanda CN: Behecet's syndrome, J Ind Med Assoc 43 : 183, 1964.
10. Pallis CA, and Fudge BA: Arch Neurol Psychiat 75 : 1, 1956. Cited by 7.
11. Planner H, and Romanovsky F: Arch Derm Syph (Berlin) 140 : 162, 1940. Cited by 7.
12. Rook A, et al: Text book of dermatology, 2nd printing, Blackwell Scientific publications, Oxford and Edinburgh, 1968, p 1479.
13. Sezer N: Further investigations on the virus of Behecet's disease, Amer J Ophthal 41 : 41, 1956.
14. Smith RB: Behecets disease with retinal vascular lesions Brit Med J 2 : 220, 1967.
15. Strancher RW, and Wigzell FW: Ann Rheum Dis 22 : 26, 1963. Cited by 7.
16. Thomas EWP: Brit Med J 1 : 14, 1947. Cited by 7.

TRUE or FALSE?

All patients with atopic dermatitis have a raised serum level of immunoglobulin E which increases with activity of the clinical condition and decreases when the dermatitis subsides.

(Answer page No. 217)