Short Case Reports

Behçet's disease

Alleviation of buccal and genital ulceration by an oral contraceptive agent

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Since Behçet's first description in 1937 of the triad of aphthous stomatitis, genital ulcers, and recurrent uveitis, many examples of *formes frustes* have been described (Curth, 1946).

The cause of the disorder is unknown, but Behçet (1939) suggested that a virus was responsible and more recently the mycoplasmas have been implicated, but without any very convincing evidence. The essential pathology in the condition is a granulomatous phlebitis (Hartemann, Schmitt, Tridon, Maken, Floquet and Guazzi, 1966) or vasculitis (Cunliffe and Menon, 1969) and the postulation of an autoimmune causative mechanism (Lehner, 1967) is attractive.

Bernstein and Suarez (1963) reported an initially monosymptomatic case of Behçet's disease in a young girl, in whom the condition began as recurring aphthae of the oral mucosa which evolved explosively into the classical triple symptom complex. The disorder had previously exacerbated regularly with the menses and this similarity to dermatitis dysmenorrhoeica (which is frequently relieved by oestrogen administration) caused them to agree with Shelley, Preucel, and Spoot (1964) that the reaction pattern represented an autoimmune vasculitis wherein this female patient had become sensitized to her own progesterone. They suppressed the endogenous progesterone production with gratifying results on the disease process.

The case presented here shows certain similarities to that of Bernstein and Suarez.

Case report

A 19-year-old single English girl first attended the West London Hospital in April, 1968; she complained of vulval ulceration which had been present for one week and was becoming worse. She had never had sexual intercourse. She gave a history of buccal ulceration recurring at about the time of menstruation for some years, but she had never had any previous genital trouble.

EXAMINATION

She was generally fit and slightly built.

Mouth There was a craggy ulcer $\frac{1}{4}$ " diameter on the right buccal mucosa (Figure). The tongue and cheeks showed considerable scarring from previous ulcers.

FIGURE *Appearance of buccal ulceration*

Genitalia There was a craggy ulcer $1\frac{1}{2}$ " by $\frac{3}{4}$ " on the right labium majus and tender, enlarged inguinal lymph nodes. Otherwise no abnormality.

Other systems No abnormality detected.

INVESTIGATIONS

Cardiolipin Wassermann reaction Negative

V.D.R.L. slide test Negative

Reiter protein complement-fixation test Negative

Urethral and cervical smears and cultures Negative for *N. gonorrhoeae*

Wet vaginal smear No trichomonads; no *Candida*

Darkground examination of serum from vulval ulcer Negative for *T. pallidum*

Haemoglobin 96 per cent. 14.0 g. per cent.

Packed cell volume 42 per cent.

Mean corpuscular haemoglobin concentration 34 per cent.

White blood cells 8,100 per cubic mm.

Neutrophils 69 per cent.

Lymphocytes 28 per cent.

Eosinophils 1 per cent.

Monocytes 2 per cent.

Erythrocyte sedimentation rate 16 mm./hr (Westergren)
Bacteriology of swab from vulval ulcer No acid-fast
 bacilli. Scanty growth of beta-haemolytic streptococci
 not of Lancefield group A.
Mantoux test 1 in 10,000: Negative

TREATMENT AND PROGRESS

When it became obvious that outpatient treatment was unlikely to succeed, she was admitted on the following day. She was treated initially with procaine penicillin, and as she failed to improve she was given tetracycline 250 mg. four times a day; she was discharged 4 days later on this treatment.

Comment

Since this episode of severe ulceration her condition has waxed and waned but none of the exacerbations has been as bad as the first.

Hydrocortisone hemisuccinate pellets made the buccal condition worse, and other forms of local treatment proved unhelpful. Tetracycline by mouth appeared to alleviate the condition but failed to produce any dramatic improvement. Since May, 1969, the patient has been treated with 'C-Quens', an oral contraceptive agent which consists of Mestranol 0.1 mg. given for 14 days, followed by a 7-day course of Mestranol 0.1 mg. with Chlormadione 1.5 mg., followed by 7 days without tablets before starting the next course. Her condition has improved while she has been taking this contraceptive regime; it is suggested that her condition may be due to sensitization to her own progesterone so that the partial suppression of endogenous progesterone production caused by the pill has had a beneficial effect on the disease process.

Differential diagnosis

The appearance is suggestive of the 'Periadenitis mucosa necrotica recurrens' described by Sutton

(1911), but the genital involvement and the poor response to Corlan (hydrocortisone) would seem to militate against this.

The history of the disease and its gratifying response to hormonal therapy is more suggestive of cyclic buccal ulceration, but the appearance of the lesions is not typical and I would favour a *forme fruste* of Behçet's disease as the most likely diagnosis.

Summary

In the case of a *forme fruste* of Behçet's disease, buccal and genital ulceration recurring with menstruation was alleviated by the use of an oral contraceptive agent.

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Maladie de Behçet

Amélioration de l'ulcération des régions buccale et génitale par la prise d'un contraceptif oral

RÉSUMÉ

Dans un cas de forme fruste de maladie de Behçet, l'ulcération des régions buccale et génitale, qui revenait avec les règles, fut améliorée et soulagée par la prise d'un contraceptif oral.