

Behçet's disease with recurrent facial paralysis

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Behçet's disease is characterized by hypopyon iridocyclitis, oro-genital ulceration, arthropathy, thrombophlebitis, and various neurological complications. The disease usually runs a relatively benign course, manifested by intermittent episodes of mucocutaneous and ocular symptoms lasting from days to weeks.

Case report

A female patient now aged 32 first attended the out-patients department at the Norfolk and Norwich Hospital in 1955 at the age of 15 with right Bell's palsy, which was treated by physiotherapy. There was a previous history of right Bell's palsy in 1949. She attended again in 1956 with right Bell's palsy which cleared without any specific treatment in about 4 weeks. X ray of the mastoid bone showed no abnormality.

In 1963 she developed vaginal discharge after the birth of her first baby. The discharge did not improve despite treatment. In October, 1965, she complained of moving spots in front of the left eye. Visual acuity in both eyes was 6/6. A diagnosis of left uveitis was made. No precipitating cause was found.

Routine haematological and radiological investigations were negative. Scrapings from the mouth ulcer (Figure) and the conjunctiva showed no inclusion bodies. The Kveim test was also negative.



FIGURE *Ulcer on inner surface of lip*

The patient was treated with topical steroids and mydriatics but did not improve. Whilst she was under treatment for uveitis, she started having episodes of high fever, pain, and stiffness of the joints. In April, 1966, she also complained of mouth ulcers and recalled having had them previously. A clinical diagnosis of Behçet's disease was established. She was started on a small dose of prednisolone, 5 mg. three times a day, and the uveitis started to improve but did not settle completely.

In May, 1967, she developed secondary glaucoma which was kept under control with Diamox. Systemic and topical steroids were stopped for a while as it was thought these might be the precipitating factor. As the uveitis did not settle, prednisolone 5 mg. three times a day was restarted. In May, 1968, she had an episode of haematemesis, while still on prednisolone. Barium meal swallow and follow-through showed no abnormality. In view of the haematemesis the dose of prednisolone was reduced. In July, 1968, she again developed secondary glaucoma. Steroids both topical and systemic were discontinued as the uveitis had settled. The intraocular pressure was controlled by Diamox.

The visual acuity in the left eye was reduced to 6/24 because of the slow development of posterior capsular lens opacities, whereas it was 6/6 in the right eye. Phenylbutazone 100 mg. twice a day was prescribed for the joint pains. In July, 1969, and June, 1970, she had a recurrence of right Bell's palsy, and in February, 1971, she had left Bell's palsy for the first time. These palsies both improved with a course of ACTH.

CHARACTERISTICS OF FACIAL PARALYSIS

- (a) Pyrexia for a few days.
- (b) Pain around the ear.
- (c) "Tickling" and twitching of the side of the face.
- (d) Loss of sensation of taste in the anterior two-thirds of the tongue.
- (e) Paralysis of the side of the face about a week after the onset of temperature.
- (f) Facial paralysis improved with a course of ACTH in 2 to 3 weeks.

Discussion

In this particular case it seems that the disease process started in 1949 at the age of 9, when the patient first developed right Bell's palsy which recurred in 1955 and 1956 and improved without any specific treatment. The slow rate of visual loss can be ascribed to the early and better control achieved with steroids. The fact that the recurrences of Bell's palsy in 1969, 1970, and 1971 improved with the use of ACTH favours an allergic cause and there seems to be no doubt that steroids do reduce the severity of the disease. Haematemesis could just be a coincidence or the result of a gastric ulcer aggravated by systemic steroids, and the absence of gastric ulcer on barium meal swallow and follow-through does not rule out the possibility that the ulcer might have healed like the mouth ulcers before the barium meal was carried out.

Summary

A case is reported of Behçet's disease with an unusual combination of haematemesis and recurrent facial paralysis. The condition improved with ACTH.

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