fever or acute rheumatoid arthritis, and stresses its brief duration in the tuberculous cases. The case reported here would appear to fulfil these criteria. It is interesting to recall in this connexion the well-authenticated syndrome of erythema nodosum, arthralgia, and hilar lymph-node enlargement, which is a common presentation of sarcoidosis (James *et al.*, 1956). The association between pleurisy and "tuberculous rheumatism" has been described by Poncet (1902) and Schneider (1903). As in the case of tuberculous rheumatism itself, it is not possible to evaluate many of these older reports in the light of the stricter diagnostic concepts of to-day.

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Case of Behçet's Syndrome with Neurological Involvement

The association of relapsing iritis, oral ulceration, and genital ulceration has been known for some time, and in 1937 Behçet suggested that the triad forms a specific disease. It is now recognized that the condition has wider manifestations and that these may include skin lesions, arthralgia, thrombophlebitis, and neurological lesions. The case reported here is another example of Behçet's syndrome with neurological involvement.

CASE REPORT

The patient was a married woman aged 42 with two grown-up children. Her past history included operative repair of a right convergent strabismus, which had been present since birth, and an isolated attack of thrombophlebitis of the left leg in 1950. In the middle of March, 1956, she developed severe, persistent pain in the left shoulder region. She attributed the pain to a heavy fall on the left shoulder, but there had been an interval of several days between the fall and the onset of the pain. X-ray examination of the shoulder was negative and she was given a course of physiotherapy at another hospital, but without much relief. Two weeks later she noticed dysuria and soreness of her vulva on walking, and discovered an ulcer about the size of a penny on that organ. This was followed by the development of small, slightly painful ulcers on the mucous membranes of her lips. A few days later, whilst out walking, her legs felt very weak and she had to be assisted home. The right leg improved overnight, but paralysis of her left leg persisted, and on the following day she noticed that her left arm also was paralysed. At this stage, April 16, she was admitted to Paddington General Hospital.

Examination showed a pale, middle-aged woman who was co-operative but rather depressed. She had a left hemiparesis of severe degree with increased deep reflexes on the left, an extensor plantar response, and absent left abdominal reflexes. There was no sensory loss. Two small ulcers were seen on the mucous membrane of her lower lip, and on the right labium minorum there was a shallow, circular ulcer about $\frac{1}{2}$ in. (1.3 cm.) in diameter with a granulating base. There were no palpable inguinal glands. Her blood pressure was 145/90. The C.S.F. findings were: normal pressure; lymphocytes, 64/c.mm.; protein, 60 mg./100 ml.; no excess of globulin; chlorides, 720 mg./100 ml.; and negative Wassermann and Kahn reactions.

Over the next few days her symptoms and signs progressed and she complained of dizziness, blurring of vision, difficulty in swallowing, and a numb feeling over her tongue, lips, and nose. Examination on April 25 showed gross horizontal nystagmus on lateral fixation and spontaneous vertical nystagmus, more rapid on elevating the eyes. Both corneal

reflexes were diminished, particularly on the left, and there was slight diminution of pain sense over the right forehead. Bilateral facial weakness was present, more marked in the upper face on the left and in the lower face on the right. Taste was defective over the anterior two-thirds of the tongue. There was right palatal weakness. She had a feeble cough and dysarthria which appeared to be a combination of pharyngeal weakness and cerebellar disturbance. There was weakness of the left trapezius and sternomastoid. Her left hemiparesis remained unchanged.

In view of the rapidly developing bulbar signs the patient was transferred to the respiratory unit at the National Hospital for Nervous Diseases, Queen Square. A repeat examination of the C.S.F. performed there showed a fall in the lymphocyte count to 13/c.mm. and a slight increase in protein to 95 mg./100 ml. No further deterioration in her condition occurred and the use of a respirator was not required. She was kept under observation and her brain-stem signs diminished considerably, her hemiparesis improved, and the ulcer on her vulva healed. When she was transferred back to Paddington General Hospital on May 11, she was able to walk, but the weakness of her left arm remained severe. The only abnormalities of her cranial nerves were a slight left lower facial weakness and slight nystagmus on extreme lateral fixation.

She was discharged on June 7, and the recovery from her hemiparesis has continued. She has since had further ulcers on her lips and buccal mucosa from time to time, but no further genital ulceration. In October she described several small skin lesions which appeared on her left arm, left shin, and right groin. These apparently started as vesicles which broke down and left small ulcers which persisted for a few days. There has never been any abnormality of her eyes.

COMMENT

Although only two of the features of the "triple syndrome" complex are present this is accepted as being sufficient for the diagnosis of Behçet's syndrome (Curth, 1946; Phillips and Scott, 1955). Eye involvement may not occur until as long as 10 years after the initial manifestations.

Neurological complications usually occur between the second and fifth years of the disease, and only in the first case of Wadia and Williams (1957) has the history been as short as three months. The illness here reported appeared to have an acute onset, with pain in the region of the left shoulder-joint, followed by orogenital ulceration and leading in four weeks to severe neurological disorder.

As in many of the other reported cases, this patient's signs were those of a mid-brain lesion. Her case is of particular interest in that she showed sensory changes and cerebellar signs, both of which appear to be uncommon in Behçet's syndrome. There seems to have been involvement of the cerebellar tracts in the mid-brain, the sensory nuclei of the trigeminal nerve, and the facial nuclei. An interesting finding was the defective taste sensibility, pointing probably to a peripheral nerve lesion.

Pallis and Fudge (1956) pointed out that the neurological disorder usually conforms to one of the following patterns: (1) a brain-stem syndrome, (2) a meningomyelitic syndrome, and (3) an organic confusional syndrome. This classification was based on the 11 cases previously recorded and two of their own, but as further cases have been reported (McMenemey and Lawrence, 1957; Evans, Pallis, and Spillane, 1957) it has become apparent that this classification is too rigid and that no case conforms entirely to one pattern.

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