Medical Memoranda

Acute Sarcoid Meningo-encephalitis

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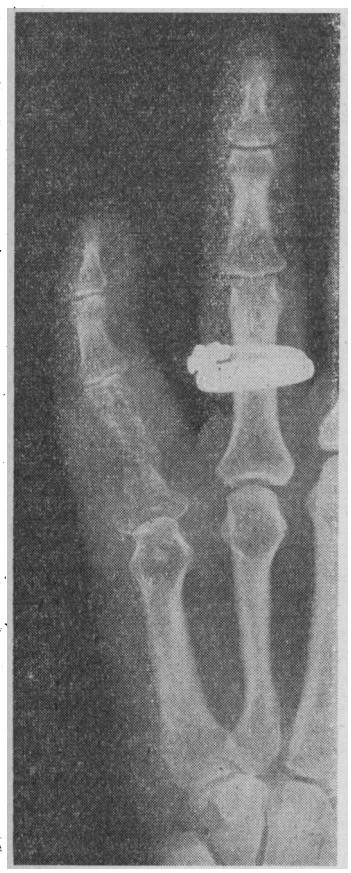
Meningo-encephalitis is a rare complication of sarcoidosis. The onset may be abrupt and deterioration rapid. Failure to be aware of this may result in the death of a patient with a treatable condition.

Case Report

A woman aged 42 was admitted to hospital on 22 January 1963 having had several generalized convulsions early that morning. There was no family history of epilepsy. The only information available on admission was that her weight had fallen by 35 kg. in two years and that she had been drowsy the previous day.



Cystic and destructive changes in the phalanges, January 1963.

She was drowsy, disorientated, and incontinent of urine and faeces. Though her neck was stiff, Kernig's sign was not present. Both plantar responses were extensor. Examination of sensory nerve function was unreliable on account of lack of co-operation, but subsequent examinations did not reveal any sensory loss. The optic fundi were normal and there were no cranial nerve lesions.

There were numerous purple areas of skin infiltration which were raised, irregular in outline, and varied from 1 to 6 cm. in greatest diameter. They were confined to the face, back of the chest, and arms, particularly the upper arms; the lesions tended to be symmetrical. There were two subcutaneous nodules without associated skin infiltration, one on the left forearm and the other on the anterior abdominal wall.

Large, rubbery, and discrete lymph nodes were palpable in the neck, supraclavicular fossae, axillae, epitrochlear regions, and groins. The left little finger and the right middle finger were swollen. The liver and spleen were not palpable. Her temperature was normal. The chest radiograph was normal, but films of the hands revealed destructive changes (see Fig.).

The lumbar cerebrospinal fluid contained 8 lymphocytes/c.mm., protein 250 mg./100 ml., sugar 18 mg./100 ml., and chloride 730 mEq/l. The Wassermann reaction was negative. The results of other investigations were haemoglobin 14.7 g./100 ml., erythrocyte sedimentation rate 32 mm. in one hour (Wintrobe), and total serum proteins 5.2 g./100 ml., of which 2.4 g. was albumin and 2.8 g. globulin (0.3 g., 0.7 g., 0.7 g., and 1.2 g.). The serum calcium was 8.6 mg./100 ml., inorganic phosphate 3.6 mg./100 ml., and alkaline phosphatase 8.3 King-Armstrong units/100 ml.

She became progressively more drowsy and had infrequent generalized convulsions. Biopsy of an epitrochlear node and an area of skin infiltration was performed on 25 January. Since her general condition was deteriorating rapidly and sarcoidosis was the only treatable condition compatible with the signs, she was given prednisone 40 mg. daily while the result of the biopsy was awaited. There was a dramatic improvement in her mental and physical condition within 48 hours. The histological features of both gland and skin infiltration were those of sarcoidosis. The prednisone was reduced over four weeks to a maintenance dose of 10 mg. daily, which was further reduced to 5 mg. in May 1963.

When she was readmitted in May 1963 for reassessment the skin infiltrations were contracted, wrinkled, and depressed, and were a greyish purple colour. No lymph nodes were palpable. The subcutaneous nodule on the abdominal wall had disappeared and the nodule on the left forearm was much smaller. The swelling of the fingers had subsided and a radiograph showed that recalcification of the phalanges had begun. The only neurological abnormality was that the plantar responses were extensor.

The cerebrospinal fluid was now normal: there was 1 lymphocyte/c.mm., protein 40 mg./100 ml., sugar 49 mg./100 ml., and chloride 710 mEq/l. The erythrocyte sedimentation rate was 35 mm.

In August 1963 the E.S.R. had fallen to 30 mm. in one hour. The total serum protein was 6.5 g./100 ml., of which 3.35 g. was albumin and 3.15 g. globulin; the electrophoretic pattern was normal.

Comment

The response to treatment with adrenal corticosteroids in patients with sarcoidosis of the central nervous system is unpredictable and is often disappointing. The signs and symptoms may diminish spontaneously without treatment. This patient was critically ill before treatment, the clinical response was dramatic, and the cerebrospinal fluid returned to normal. A similar response to treatment with adrenal corticosteroids was obtained in the only other case of acute sarcoid meningo-encephalitis that is recorded as having been treated in this way (Carstensen and Norviit 1953).

Neurological signs may develop when there is no evidence of sarcoid elsewhere, though six of seven patients described by Walker (1961) had at one time had demonstrable pulmonary involvement, which in one case was apparent only in a chest radiograph taken 10 years earlier.

Sarcoidosis should always be considered in the differential diagnosis of a patient with meningo-encephalitis, because the response to treatment with adrenal corticosteroids is excellent.

I am grateful to Dr. W. D. Brinton for permission to report his case.

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References

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