Clinical Reports

Cerebellar syndrome in sarcoidosis

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Summary: A 63 year old woman who presented with a cerebellar syndrome was shown to have sarcoidosis of the central nervous system, skin and liver with no pulmonary involvement.

Introduction

Sarcoidosis involves the nervous system in 5% of cases.¹ It usually affects the cranial nerves, particularly the facial. Granulomata may involve the meninges or the substance of the brain or spinal cord. Involvement of the cerebellum is rare and usually associated with other brainstem signs.^{2,3} Most cases of neurosarcoidosis reported have been neurological manifestations of established, usually pulmonary, sarcoidosis.^{1,2} We report a case presenting with a pure cerebellar syndrome with no demonstrated pulmonary involvement.

Case report

For one year a 63 year old woman had noted gradually increasing unsteadiness of gait, recurrent unexplained fever, felt tired and had lost 10 kg in weight. Over the previous 8 weeks she had noted a circular violaceous lesion on the dorsum of the right hand. She was on no drugs.

On examination she was cachectic and pale. Cardiovascular and respiratory systems were normal. The spleen was just palpable. Her gait was ataxic and she had cerebellar signs in the arms and legs, worse on the left. There was no nystagmus.

Investigations: haemoglobin 7.5 g/dl, erythrocyte sedimentation rate (ESR) (Westergren) 103 mm/hour, alkaline phosphatase 881 IU/l, gamma glutamyl transpeptidase 110 IU/l, albumin 32 g/l, angiotensin converting enzyme 52 IU/l (normal < 53). The following were all normal or negative: aspartate transaminase, bilirubin, calcium, autoantibodies including antimitochondrial, blood cultures, chest X-rays and X-rays of the hands and feet. Mantoux test at 1 in 1000. early

morning urine culture, cerebrospinal fluid microscopy and biochemistry, computerized axial tomography of the abdomen and ultrasound of the pelvis. Bone marrow showed changes consistent with anaemia of chronic disorders. Biopsy of the skin lesions on the hand showed non-caseating granulomata with multinucleate giant cells. On liver biopsy well circumscribed similar non-caseating granulomata were found in the periportal region. A Kveim test was negative. Computed tomography of the brain showed minor atrophic changes with patchy low density areas in the white matter. Nuclear magnetic resonance scan of the brain showed thickening of the frontal meninges and signals of high intensity scattered throughout the brain (Figure 1). Prednisolone 60 mg on alternate days produced a dramatic response: her energy returned, her walking became less ataxic, and her haemoglobin increased to 12.5 g/dl, her ESR fell to 28 mm/hour, and her alkaline phosphatase fell to 325 IU/l. The improvement was sustained over the next 4 months during which time she put on 10 kg.

Discussion

The diagnosis of sarcoidosis is made on finding non-caseating chronic granulomata in a number of organs and the exclusion of other causes of such granulomata.¹ This patient had a cerebellar syndrome, histologically proven granulomata in skin and liver, and multiple lesions on nuclear magnetic resonance scanning compatible with central nervous system sarcoidosis. The Kveim test is positive in only about a third of cases of chronic sarcoidosis.¹

There are three previous case reports of neurosarcoidosis producing only cerebellar signs,^{3,4,5} and in all three there were abnormal chest X-rays suggesting sarcoidosis.

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Figure 1 Nuclear magnetic resonance scans showing sagittal sections of the brain. The area of high intensity signals is arrowed.

References

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The good response to steroids highlights the need to consider sarcoidosis in the differential diagnosis of a pure chronic cerebellar syndrome even in the absence of detectable pulmonary involvement, particularly if associated with unexplained fever and anaemia.

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