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Carotid arteritis causing amaurosis fugax and ischaemic cerebrovascular events in neurosarcoidosis

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ABSTRACT

Objective: To present and review the vascular consequences of arteritis in neurosarcoidosis.

Patient and methods: neurosarcoidosis is typically an inflammatory disorder of the meninges surrounding the brain and spinal cord. Although inflammation of small and medium sized vessels is seen pathologically and vasculitis is occasionally described, a large intracerebral arteritis has not previously been reported. A few case reports exist, however, which describe the vascular consequences of large vessel compromise in the disorder. We review the literature and present a new case with novel MRI features which imply carotid arteritis.

Results: The case presented with a disorder of the carotid artery on one side leading to a series of TIAs. Inflammation of the wall of the carotid artery was seen adjacent to a granulomatous leptomeningitis. The disorder responded to immunosuppressive therapy without recurrence.

Conclusions: The imaging features suggest a granulomatous infiltration of the carotid artery wall leading to arteritis followed by disorganisation of the internal elastic lamina and fibrosis. The data provide further insight into the pathogenesis of neurological impairments in neurosarcoidosis. The MRI features of carotid arteritis in neurosarcoidosis have not previously been demonstrated.

1. Introduction

Neurological complications arise in 5% of patients with the multisystem auto-inflammatory disease sarcoidosis. Isolated cranial neuropathies are the most prevalent clinical manifestation, but any part of the nervous system may be affected. When the brain is involved, the predominate disorder is a spreading meningoencephalitis in which leptomeningeal enhancement is seen on MRI [1]; involvement of the basal meninges and adjacent structures such as the chiasm, hypothalamus and upper brainstem is the most common [1,2].

Granulomatous inflammation of blood vessels has been known since early pathological descriptions [2–8], but only a few reports with clinical consequences of vascular involvement exist, suggesting that vascular involvement in sarcoidosis rarely causes ischaemic clinical manifestations. We present a case which illustrates the disorder and review the pathological, clinical and neuroradiological literature.

2. Case report

This 47 year old Caucasian man known to have hypertension, hyperlipidaemia and type II diabetes presented with repeated episodes of left sided amaurosis fugax which lasted for seconds to 5 hours. On occasion there was a monocular scintillation and initially there was no headache. Laboratory investigations were normal. An MRI scan of brain showed scattered white matter lesions compatible with the consequences of small vessel disease, and ultrasounds of the heart and carotid vessels were normal. He was treated with aspirin 75 mg and the episodes reduced in frequency and duration. Over the next year he developed increasing headache, and then diplopia; he was found to have a right sided upper quadrantinopia and a partial left third nerve palsy.

His ESR, biochemical screening and serum ACE were normal. A CT of the thorax and abdomen revealed mediastinal and axillary lymphadenopathy and Gallium-67 scintigram revealed uptake in the lacrimal

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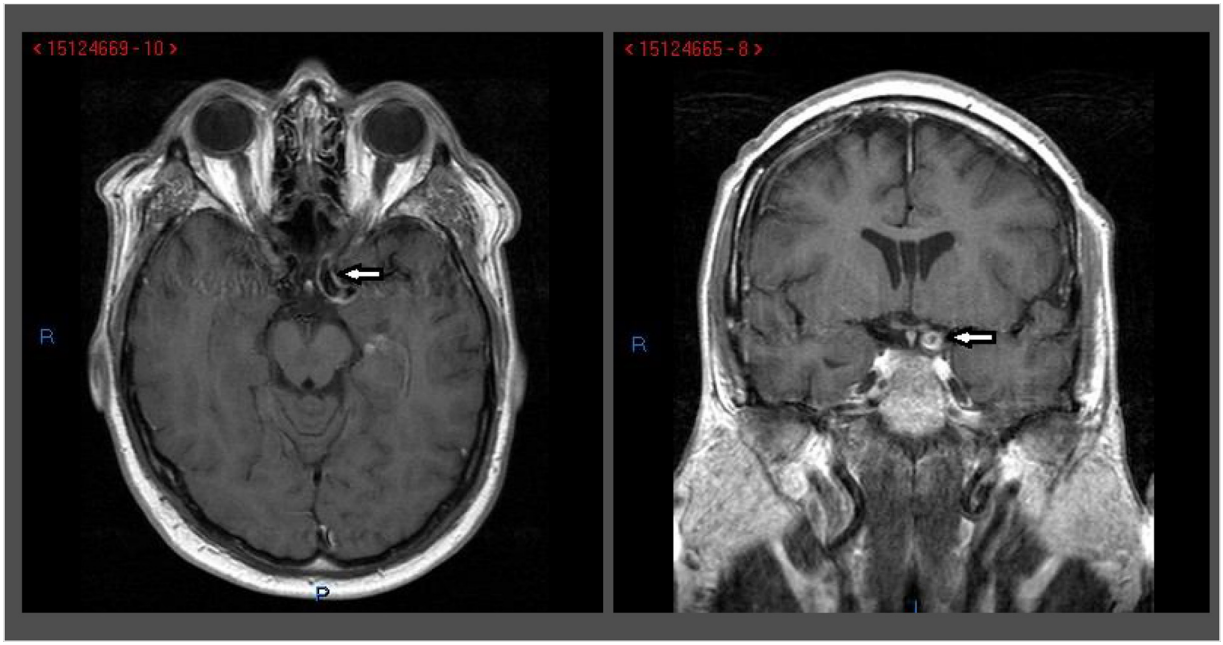

Fig. 1. (a,b): Axial and coronal T1 weighted post-contrast images with striking vessel wall enhancement along left ICA indicating acute vasculitis, with punctate cortical/subcortical enhancement in the left mesial temporal lobe relating to parenchymal disease.

and parotid glands, the mediastinal and axillary nodes and liver. A transjugular liver biopsy revealed granulomatous inflammation.

MRI of the brain (Fig. 1) showed thickening and enhancement of the left internal carotid artery, and an enhancing intrinsic lesion of the left temporal lobe with meningeal enhancement extending back to the midbrain. The middle ear clefts and mastoid air cells were opacified.

A biopsy of the temporal lesion (Fig. 2) revealed granulomatous inflammation.

He was treated with intravenous then oral corticosteroids and experienced two further episodes of amaurosis fugax as the dose was reduced below 30 mg per day. Methotrexate was added and the enhancement resolved; the carotid artery became ectatic. Thereafter he has remained well for the past five years.

3. Discussion

Herring and Urich noted that “the most constant post-mortem finding in sarcoidosis of the central nervous system is the presence of typical sarcoid granulomata in the leptomeninges and in the parenchyma of the brain” [2]. The basal leptomeninges were seen to be more frequently and more severely affected, and granulomas situated within the parenchyma had spread from the surface along the perivascular spaces of perforating blood vessels. They also noted that the walls of small and medium sized arteries were sometimes seen to have

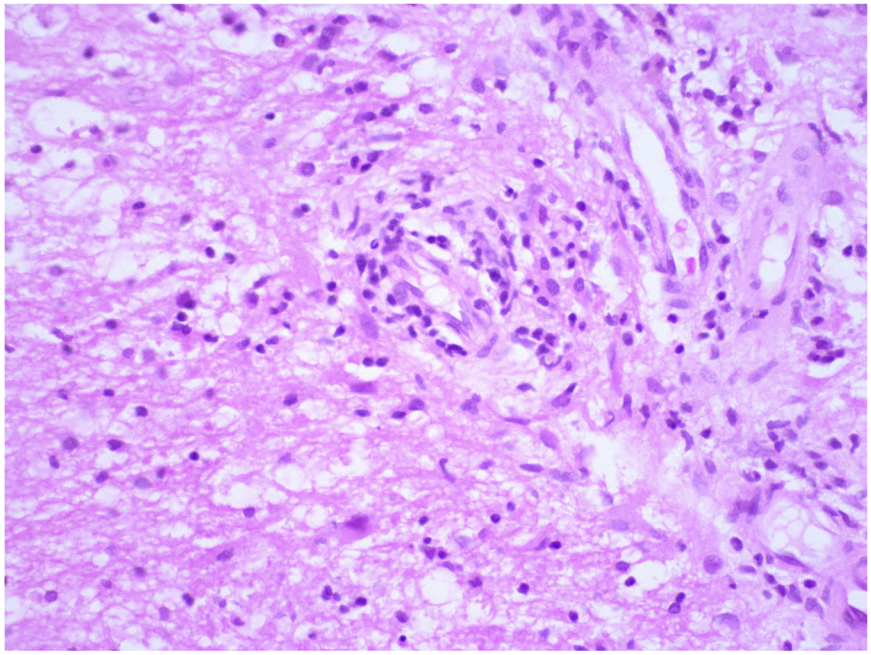

Fig. 2. Haematoxylin and eosin stained biopsy of cerebral white matter, showing gliotic white matter infiltrated by granulomas composed of epthelioid macrophages. Some are surrounded by a lymphocytic infiltrate.