Neurosarcoidosis presenting as ghost lesions in the CNS: a diagnostic dilemma

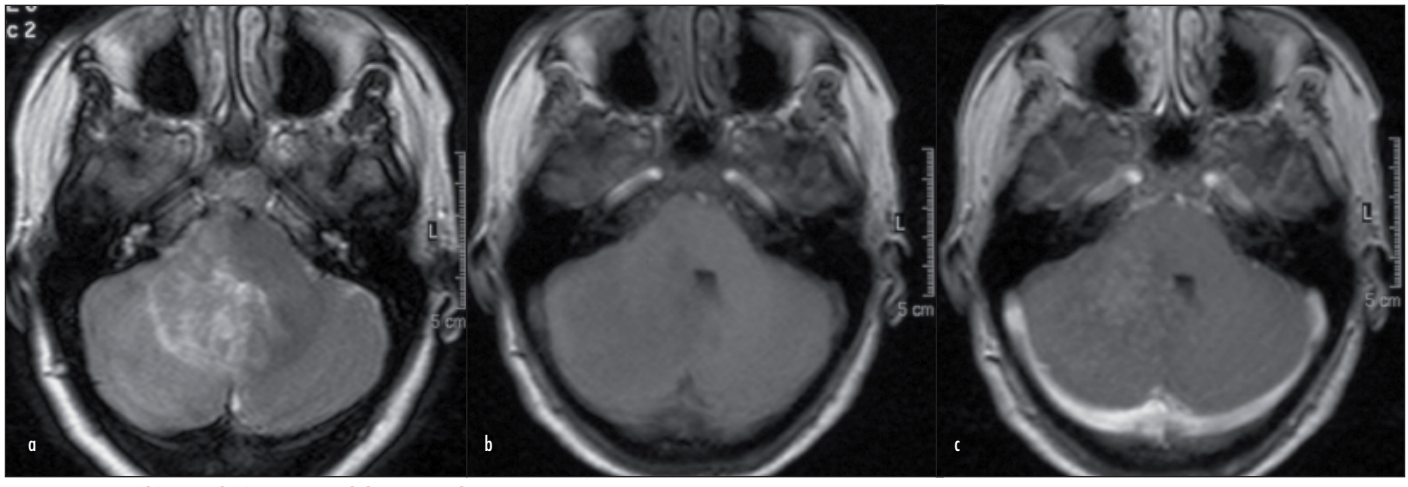

Figure 1. a. T2, (b) T1 and (c) T1 post gadolinium axial magnetic resonance imaging scan.

Introduction

This article reports an unusual case of neurosarcoidosis presenting as a cerebellar mass lesion. Resolution of the lesion with steroids prompted further searches for disease outside the CNS. Pulmonary sarcoidosis was confirmed. It also reviews the key diagnostic aspects of neurosarcoidosis presenting as CNS ghost lesions, emphasizing the importance of attempting diagnosis of atypical CNS lesions without resorting to brain biopsy.

Discussion

Radiologically, neurosarcoidosis may resemble tumours, vascular disease, demyelination, other granulomatous disease and infections (Hoitsma et al, 2004).

When seen on serial imaging as 'ghost lesions' one differential diagnosis is cerebral lymphoma. Mimicry and ghost lesions can lead to neurosarcoidosis being misdiagnosed, especially in the absence of systemic involvement. Therefore, a high index of suspicion for neurosarcoid should be maintained and diagnostic attempts made before brain biopsy, with its inherent risk of complications (Quinones-Hinojosa et al, 2003).

Since isolated neurosarcoidosis is uncommon, the presence of an unusual mass lesion should prompt an exhaustive search for extra-CNS disease even in the absence of symptoms (Kellinghaus et al, 2004). Investigations include chest X-ray

Case Report

A 50-year-old woman presented with a short history of headaches and dizziness. One week earlier she noticed impaired right leg coordination and had fallen twice. She had no significant past medical history. Examination showed impaired right arm and leg coordination, mild pronator drift and gait ataxia with no other neurological deficits.

A magnetic resonance imaging scan of the brain showed an intra-axial mass lesion centred on the right middle cerebellar peduncle with partial effacement of the fourth ventricle and an unusual, slightly nodular pattern of enhancement (*Figure 1*). A computed tomography scan of the chest demonstrated bilateral nodular hilar masses felt consistent with reactive lymphadenopathy. No single large mass was present. Brain magnetic resonance imaging was consistent with either a primary lesion or atypical metastasis.

She commenced dexamethasone 4 mg four times per day and stereotactic biopsy was scheduled. The planning magnetic resonance imaging, 2 weeks later, demonstrated marked reduction in the lesion's size raising the possibility of CNS lymphoma. Surgery was cancelled. Lumbar puncture showed no abnormalities.

The steroid dose was reduced as a result of cushingoid sequelae and to establish if the lesion would re-expand. Interval scans showed progressive reduction in mass size despite decreasing steroid doses. Partial signal normalization within the cerebellum with resolution of mass effect and enhancement was observed (*Figure 2*).

Repeat computed tomography of the chest (*Figure 3*) showed new, small, peripheral, left and right upper lobe pulmonary lesions. Histology following open biopsy of the left-sided lesion showed non-coalescent non-necrotizing granulomata consistent with sarcoidosis. Six months later the patient remains symptom free on prednisolone 2.5 mg once per day.

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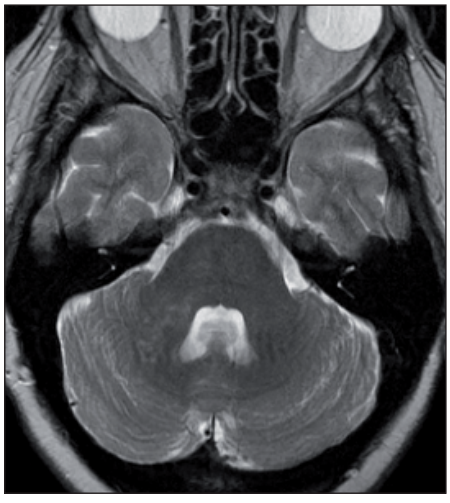
and computed tomography (90% have occult or symptomatic pulmonary involvement (Hoitsma et al, 2004), serum angiotensin-converting enzyme levels, bronchoscopy, lymph node biopsies, and CSF angiotensin-converting enzyme protein, immunoglobulins, leukocytes and trial of steroids (Uchino et al, 2001; Di Comite and Sabbadini, 2005). If surgery is not performed on intracranial masses, they require close surveillance, especially if in the posterior fossa.

Figure 2. T2 axial magnetic resonance imaging scan (3 months after* Figure 1*).



Conclusions

Given that isolated neurosarcoidosis is rare, the authors recommend exhaustive searches for evidence of extracranial involvement before brain biopsy in the context of atypical inflammatory CNS lesions. BJHM

Di Comite G, Sabbadini MG (2005) Neurological involvement in rheumatological diseases. *Neurol Sci* **26** Suppl 1: S9–14

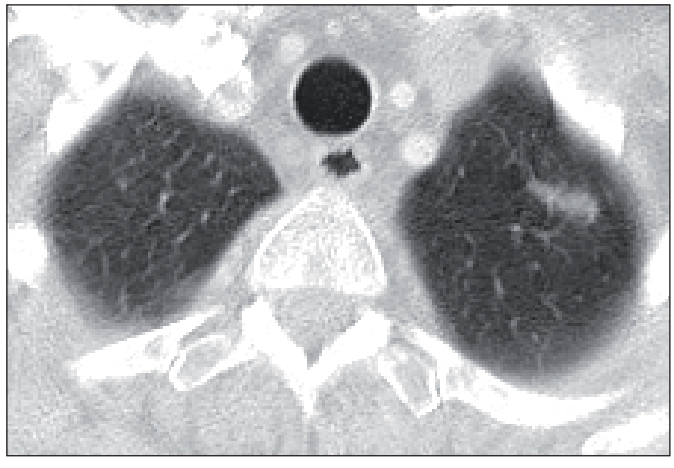
Hoitsma E, Faber CG, Drent M, Sharma OP (2004) Neurosarcoidosis: a clinical dilemma. *Lancet Neurol* **3**(7): 397–407

Kellinghaus C, Schilling M, Ludemann P (2004) Neurosarcoidosis: clinical experience and diagnostic pitfalls. *Eur Neurol* **51**(2): 84–8

Quinones-Hinojosa A, Chang EF, Khan SA, McDermott MW (2003) Isolated trigeminal nerve sarcoid granuloma mimicking trigeminal schwannoma: case report. *Neurosurgery* **52**(3): 700–5

Uchino M, Nagao T, Harada N, Shibata I, Hamatani S, Mutou H (2001) Neurosarcoidosis without systemic sarcoidosis--case report. *Neurol Med Chir (Tokyo)* **41**(1): 48–51

Figure 3. Axial computed tomography scan of the chest.



Scleral pigmentation and a heart murmur

A 73-year-old man was referred because of a heart murmur. An echocardiogram revealed moderate aortic stenosis. Since his youth he had noted that his urine turned a dark reddish-brown colour on standing. As well as pigmentation of the sclera of his eyes (*Figure 1*), the auricular cartilage of his ears also had a bluish discolouration.

The diagnosis is alkaptonuria, a rare recessive disorder (1 in 1 million births). A deficiency of the enzyme homogentisic acid oxidase leads to an accumulation of homogentisic acid. This results in blue-black discolouration of connective tissues (ochronosis), including bone, cartilage and skin. Some homogentisic acid is excreted in the urine (alkaptonuria) and forms the basis for making a diagnosis. Scleral pigmentation is evident in most patients by their forties (Phornphutkul et al, 2002).

Heart valves become pigmented and frequently degenerative. This may result in significant valvular stenosis in the elderly. Left-sided heart valves are usually involved, most frequently the aortic valve, followed by the mitral valve (Erek et al, 2004). Cardiovascular sequelae are the usual cause of death, but surprisingly life expectancy is not markedly reduced. There is currently no definitive cure for alkaptonuric ochronosis. BJHM

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Erek E, Caselman FPA, Vanermen H (2004) Cardiac ochronosis: valvular heart disease with dark green discolouration of the leaflets. *Tex Heart Inst J* **31:** 445–7

Phornphutkul C, Introne WJ, Perry MB et al (2002) Natural history of alkaptonuria. *N Engl J Med* **347:** 2111–21

Figure 1. Black scleral pigmentation is seen in both eyes of the patient.