Functional MRI as a tool for assessing chiasmal visual defect in a patient with neuromyelitis optica

Bitemporal hemianopsia is the classic visual field defect of disorders that involve the optic chiasm, caused by the involvement of the crossing nasal–retinal fibres of each optic nerve. Chiasmal visual field defects are usually categorised as extrinsic because of mechanical compression from adjacent structures, although lesions involving the substance of the chiasm itself also exist.1

We describe a unique case of chiasmal neuritis, a woman with serologically proven neuromyelitis optica (NMO).2 Our study addresses the cortical consequences of this acquired bitemporal hemianopsia. Functional MRI (fMRI) was used to evaluate the cortical activation patterns and its correspondence with visual field representation during the attack and subsequent recovery.

A 36-year-old woman presented with acute bilateral visual loss progressing during several days before her admission. Her history included serologically positive myasthenia gravis at the age of 10 years, which required immunosuppressive treatment that led to clinical remission and cessation of all drug treatment at age 13 years. She was asymptomatic until age 26 years, when she presented with an episode of transverse myelitis. Since then, she has had another three severe myelitic episodes, without optic nerve involvement and was, therefore, under maintained treatment of prednisone and cyclophosphamide. Six months before her current admission, she was tested positive for NMO immunoglobulin.2 To note is that her daily cyclophosphamide dosage was reduced just before her current admission.

Figure 1 (A) Visual fields during the acute phase (left) and subsequent recovery (right). (B) Coronal T1-weighted MRI during the acute phase (the optic chiasm is shown in the blow-up view).

Figure 2 (A) Experimental design. Four experimental conditions were used: (1) stimuli projected to the LVF via the LE, marked in yellow; (2) stimuli projected to the RVF via the LE, marked in light green; (3) stimuli projected to the LVF via the RE, marked in red; (4) stimuli projected to the RVF via the RE, marked in dark green. LE, left eye; LVF, left visual field; RE, right eye; RVF, right visual field. (B) Coronal and axial views of the fMRI activation patterns, elicited during the four experimental conditions. Activation is shown for the posterior occipital cortex for a control participant (left) and for the patient during acute phase (middle) and after recovery (right panel). Same colours as in C (see scale). Conv. (convergence) refers to the overlap of activation elicited by stimuli projected to both eyes. (C) Average activation level (per cent signal change) histograms. Activation levels during stimuli presentation in V1, showing the relative contribution of all experimental conditions (see Supplementary Methods for details). The asterisks denote significance level, paired t test: LH P < 0.05; RH P < 0.01. (D) Crossing ratio. Assessment of the relative contribution of the ipsilateral and contralateral eyes for activating V1 in each hemisphere (see Supplementary Methods for details).
Her clinical examination revealed a parapiramidal syndrome with sensory level at T5 and mild left optic disc temporal pallor. No relative afferent pupillary defect was detected. Snellen visual acuity measurements were 6/60 in the right eye and 6/9 in the left eye. The Humphrey perimetry tests were 6/60 in the right eye and 6/9 detected. Snellen visual acuity measurements were performed in a 3 T Siemens Trio scanner. Notopic mapping was performed. The fMRI results can be observed in the cortical activation of the retinogeniculate pathway. fMRI had a major impact on cognitive neuroscience, and its ability to demonstrate a function-structure relationship in healthy volunteers has been demonstrated clearly in the visual system. However, it is still in its earliest stages of transition from research laboratories to clinical application.

The cortical consequence of chiasmal abnormalities had previously been limited to congenital abnormalities as in an achiasmatic patient, in whom each eye projects to the ipsilateral hemisphere, and in albinos, in whom most of the retinal ganglion cells cross to the contralateral side. Our study, however, described the cortical consequences of acquired (adult) bitemporal hemianopsia, in which the nasal–retinal fibres are temporarily lost rather than misrouted. We demonstrated the outcome of damage and recovery to the optic chiasm formation.

This study illustrates the benefit of adding fMRI to standard anatomical scans. We, therefore, recommend using fMRI as a clinical tool for studying the neurological basis of visual field impairments.

Activated microglia in the subthalamic nucleus in hyperglycaemic hemiballism: a case report

Non-ketotic hyperglycaemia is one of the causes of hemiballism. The involuntary movement is also sometimes referred to as hemichorea. Hyperglycaemic hemiballism has been thought to occur in patients with poorly controlled diabetes. MRI usually shows a hyperintense lesion of the contralateral basal ganglia in T2-weighted image. Although there have been a few pathological studies of hyperglycaemic hemiballism, no studies have shown pathological alteration in the subthalamic nucleus (corpus Luysii) which is responsible for the hemiballism resulting from ischaemia. Originally, Schwarz and Barrows reported hyperglycaemic hemiballism as ‘hemiballism without involvement of Luys’ body.’ We herein report a case of hyperglycaemic hemiballism in which immunohistochemistry showed the presence of activated microglia in the subthalamic nucleus contra-lateral to the hemiballism.