Dyke–Davidoff–Masson Syndrome Demonstrated by Current MR images
A Case Report

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SUMMARY – *Current magnetic resonance imaging techniques demonstrated MR findings of Dyke–Davidoff–Masson syndrome in a 44-year-old man. Statistical parametric mapping analysis of the T1-weighted images showed focal atrophy in the basal ganglia. Three-dimensional white matter fibers of corticospinal tracts, corpus callosum and cingulate bundle were demonstrated using diffusion tensor data correlated to the patient's clinical conditions.*

Introduction

Dyke–Davidoff–Masson syndrome (DDMS) is an uncommon disease characterized by cerebral hemiatrophy accompanied by ipsilateral compensatory skull and sinus changes [1-5].

Imaging findings of DDMS have been described on computed tomography (CT) and conventional magnetic resonance imaging (MRI) including T1- or T2-weighted imaging in the literature [2-5].

Nowadays diffusion tensor MRI (DTI) and its functional maps, including fractional anisotropy and three-dimensional white matter fiber tracking maps, are used to evaluate brain structure and disorder.

In the past, only one case report of this disease with fractional anisotropy map was reported in the literature [5].

This paper analyzes and describes the findings of 3D white matter tractography in a case of DDMS and correlates the findings with the patient's clinical symptoms.

To our knowledge, this is the first case report of DDMS on 3D white matter tractography using diffusion tensor MRI data.

Case Report

A 44-year-old man previously diagnosed with a DDMS who suffered a recent seizure was referred to our institution for further evaluation. The patient had been diagnosed with schizophrenia and also mild mental retardation. Physical examination revealed no motor disorder. All MR imaging was performed on a 1.5 Tesla MR system (EXCELART Vantage XGV1.5T,

Toshiba Medical Systems, Tokyo, Japan) with a five-channel head coil at our institution. Brain MR imaging protocol consisted of T1-weighted 3D gradient echo in sagittal plane (TR/TE/FA; 24.4 ms/5.5 ms/35°, 256×256 matrix, 1 mm section thickness), T2-weighted fast spin- echo (TR/TE; 8800 ms/107 ms, 256×192 matrix, 2 mm section thickness), and echo-planar diffusion tensor imaging (TR/TE; 7668 ms/100 ms, 128×128 matrix, 5mm section thickness) in axial plane. All diffusion tensor images were transferred to an independent workstation and calculated. Mean fractional anisotropy maps on gray-scale and three dimensional tractography of white matter fibers

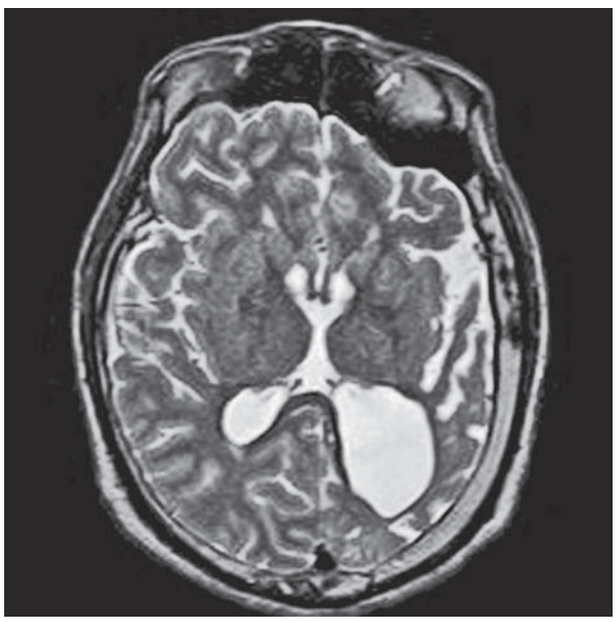


Figure 1 Axial T2-weighted image shows the left hemiatrophy and mild changes in the ipsilateral basal ganglia.

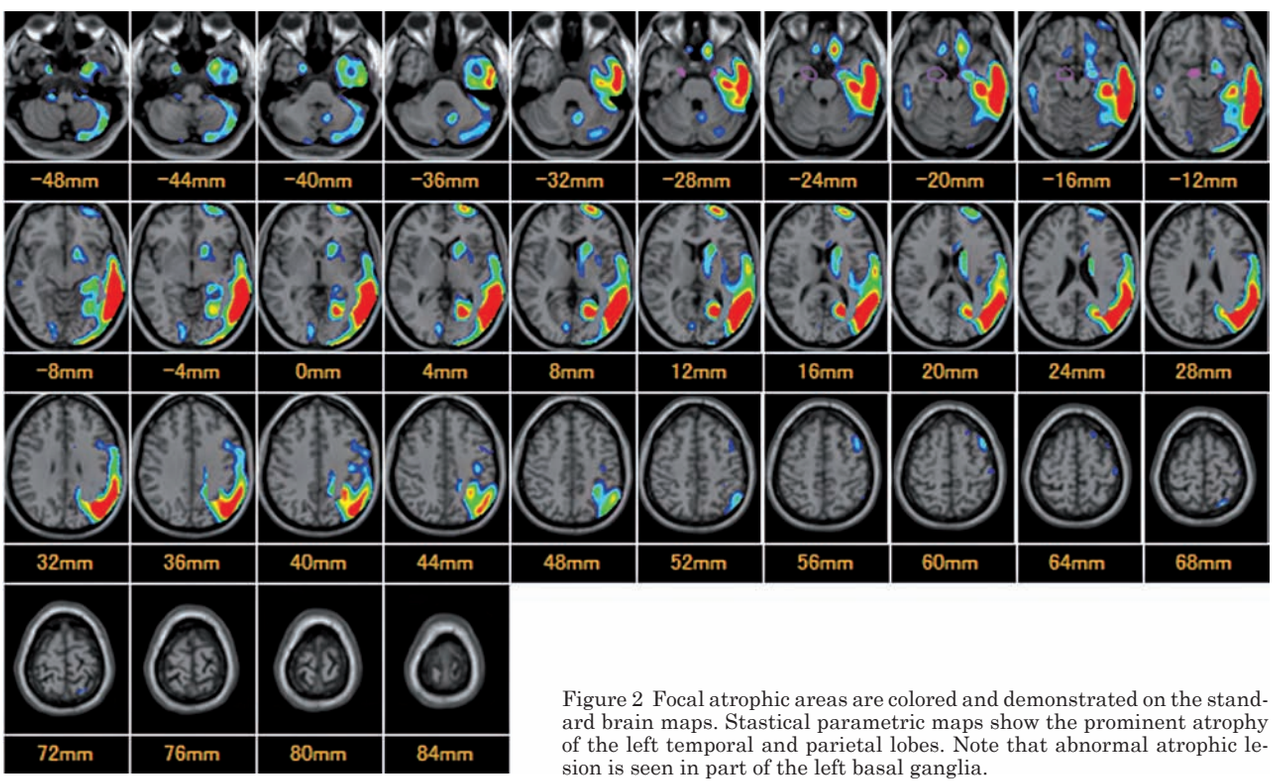
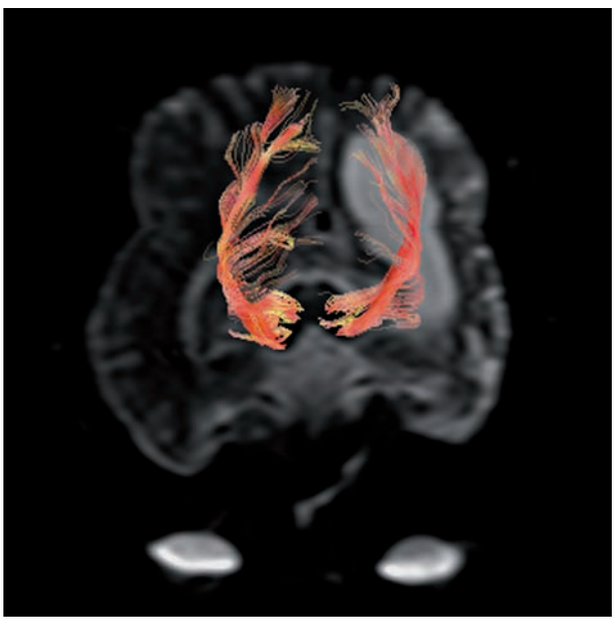


Figure 2 Focal atrophic areas are colored and demonstrated on the standard brain maps. Stastical parametric maps show the prominent atrophy of the left temporal and parietal lobes. Note that abnormal atrophic lesion is seen in part of the left basal ganglia.

consisting of three major fibers, corticospinal tracts, corpus callosum and cingulate bundle fibers were obtained by using software (dTVII, developed by Image Computing and Analysis Laboratory, Tokyo, Japan). For the three-dimensional reconstruction of white matter fibers, we used the region-of-interest (ROI) approach [6,7].

MRI revealed left cerebral hemiatrophy without midline shift on the T1-weighted and T2-weighted images (figure 1) and the left basal ganglia showed mild abnormal findings.



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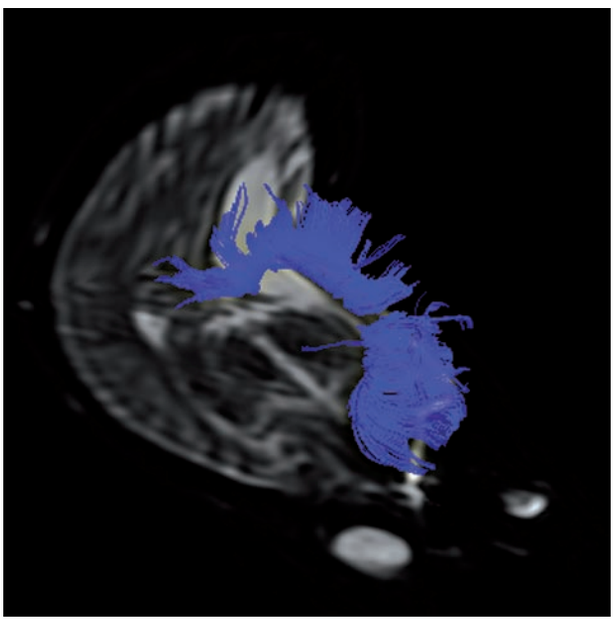

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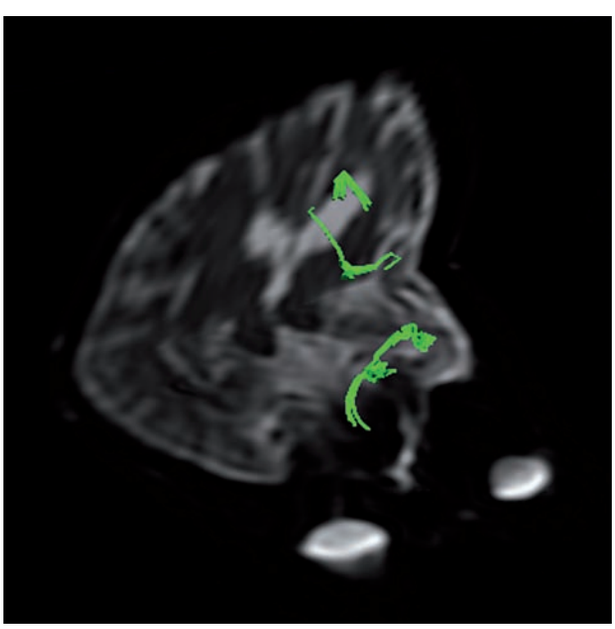

C

Figure 3 Three-dimensional tractography of white matter fibers consists of corticospinal tracts (A) and corpus callosum (B) are normally demonstrated but the fibers of cingulate bundle (C) are poorly demonstrated.

However, statistical parametric mapping analysis of the T1-weighted images using software VSRAD [8] showed focal atrophy in the basal ganglia (figure 2). As results of white matter tractography, bilateral corticospinal tracts and corpus callosum were demonstrated almost equally (figure 3A,B). The demonstration of the fibers agreed with the clinical findings that this patient showed no motor disturbance. On the contrary, poorly demonstrated cingulate bundle fibers were thought to reflect long-term schizophrenia (figure 3C).

Discussion

Diagnosis of DDMS is straightforward, even when CT or MR images of the patient are not available. Characteristic appearance and only a plane radiograph of the skull will establish the diagnosis. In fact, CT or MR images did not exist when this disease was first described in 1933 by Dyke, Davidoff and Masson [1]. However, nowadays, the diagnosis of DDMS means little because it is ascribed to patients with cerebral hemiatrophy. There is a wide variety of the brain damage including basal ganglia, causes (congenital or acquired), etiology and symptoms of this DDMS. To comprehend the degree of the patient with recent MR imaging techniques, more detailed investigation is needed.

In our case, small changes in atrophy and abnormal intensity of the basal ganglia were seen on T1- and T2-weighted images but significant atrophy was displayed by statistical parametric mapping analysis. Moreover, the atrophy was not seen in the internal capsule. This was important because the patient showed no motor disturbance. In addition to this finding, 3D tractography of corticospinal tracts and corpus callosum were normal and also supported the patient's conditions. By contrast, cingulum bundle abnormality was detected because of schizophrenia as reported in the literature [9,10]. Therefore, statistical parametric mapping analysis and 3D tractography showed more useful information for this patient, although they seem to exceed the usual MR examination.

Conclusion

We described MRI findings in a case of DDMS using thin slice T1-weighted imaging and DTI. Recent MR imaging sequences such as 3D white matter tractography are useful not only for the diagnosis of this disease, but also to comprehend patients' clinical conditions.

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