Case description

A 26-year-old right-handed woman with no history of neurological illness was referred to the department of clinical neurology of our hospital for evaluation of her continuous involuntary movements of 6 days’ duration. About 4 months prior to this evaluation, a transient episode of abnormal involuntary movements had developed in association with acute pharyngitis and fever lasting 2 days. The movement disorders in that episode were similar to those of this, more recent, event.

On admission, the patient underwent brain magnetic resonance imaging (MRI) and computed tomography (CT), both of which were unremarkable. She showed severe emotional lability and atypical, gradually worsening, choreic movements, consisting of irregular, unpredictable, flowing and large-amplitude actions involving the left hemibody and face. A mild diffuse hypotonia was noted, with extensor plantar reflexes, and the deep tendon reflexes were mildly decreased. Global cognitive function assessment, carried out according to a standardized battery of tests including the Mini-Mental State Examination (MMSE) and other neuropsychological tests, was normal.

Also at the time of admission, laboratory studies revealed an antistreptolysin-O titer of 531 IU/mL (normal: < 200 IU/mL), a C-reactive protein of 7.27 mg/dL (normal: < 5 mg/dL) and an erythrocyte sedimentation rate (ESR) of 24 mm/h. The results of other routine blood chemistry tests, such as for antiphospholipid antibody, lupus anticoagulant antibody, serum copper and ceruloplasmin, as well as tests for thyroid function, rheumatic factor, and antinuclear and Mycoplasma antibody, were all normal. Throat culture was positive for Streptococcus pyogenes (group A).

18F-Fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) showed bilateral basal ganglia hypermetabolism that was more prominent in the right striatum (Fig. 1). The diagnosis of Sydenham chorea (SC) was made, and treatment with haloperidol and penicillin was started on the fourth day and continued for 10 days. Five days later, the patient’s choreic symptoms began to gradually diminish, with complete remission within 2 months. Two years later, the patient underwent a control 18F-FDG PET/CT, which was unremarkable.

Discussion

SC has no specific biological markers or other objective findings, which means that its diagnosis depends solely on the clinical manifestations of acute chorea and the absence of any other underlying cause. Although other manifestations of rheumatic fever may strongly support a diagnosis of SC, their presence is not mandatory, according to the modified Jones criteria [1]. There are only a few reports of brain PET studies in SC, and the majority involve pediatric patients. Ho [2] described symmetrical basal ganglia hypermetabolism in an 11-year-old girl, while Aron [3] evaluated two pediatric cases in the acute phase and during recovery, and demonstrated the full reversibility of bilateral basal ganglia hypermetabolism. To our knowledge, there has been only one case description of adult-onset SC in the literature [4], although it evaluated brain metabolism only during the acute phase.

In our present case, serial PET/CT images and semiquantitative analysis (statistical parametric mapping, SPM) clearly demonstrated reversible bilateral hypermetabolism that was more prominent in the right striatum, suggesting that increased glucose metabolism of the basal ganglia was related to the genesis of chorea, as has been observed in other studies [2,3,5].

To our knowledge, this is also the first case to correlate two sequential PET studies in an adult patient. Striatal hypermetabolism in our case may have been either the direct consequence of an underlying pathogenesis or an autoimmune-mediated inflammatory process leading to enhanced corticostriatal synaptic activity. In other autoimmune choreas, such as primary antiphospholipid syndrome and systemic lupus erythematosus, unilateral striatal hypermetabolism has been described [6] whereas, in degenerative choreas such as Huntington’s disease, hypometabolism has been shown [7]. One possible explanation is that direct neuronal striatal activation by antibodies—similar to the activation of thyroid tissue by thyroid-stimulating immunoglobulins—has occurred. Although there is no natural precedent for direct immunoglobulin-mediated neuronal activation, antibodies able to stimulate glutamate receptors have been created in experimental models [8]. Furthermore, the absence of hypermetabolic basal ganglia findings
Figure 1  Transaxial image on 18F-FDG PET during the acute phase (A); and the same slice of acute-phase PET/CT on semiquantitative analysis (statistical parametric mapping, SPM) (B), shows the area of hypermetabolism in colour (analysis: FWE (family-wise error) rate: 0.05; voxel threshold: 50 voxels). Transaxial image on 18F-FDG PET during recovery (C), and the same slice of recovery PET/CT on SPM analysis (D); note that the previously highlighted hypermetabolism has disappeared (analysis: FWE rate: 0.05; voxel threshold: 50 voxels).

during recovery could suggest that there are no long-term, permanent, neurological sequelae.

The present case shows that 18F-FDG PET/CT can be used to trace functional changes in basal-ganglia circuitry and help to increase our understanding of the non-degenerative causes of chorea.

Conflict of interest statement

The authors disclosure of any personal or financial support or author involvement with organization(s) with financial interest in the subject matter — or any actual or potential conflict of interest.

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References


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