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< 200 IU/mL), a C-reactive protein of 7.27 mg/dL (normal:

revealed an antistreptolysin-O titer of 531 IU/mL (normal:

psychological tests, was normal.

declined. Global cognitive function assessment, carried

plantar reflexes, and the deep tendon reflexes were mildly

large-amplitude actions involving the left hemibody and

ments, consisting of irregular, unpredictable, flowing and

lability and atypical, gradually worsening, choreic move-

recent, event.

On admission, the patient underwent brain magnetic res-

donant imaging (MRI) and computed tomography (CT), both

of which were unremarkable. She showed severe emotional

ly and atypical, gradually worsening, choreic move-

ments, 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 neu-

ropsychological tests, was normal.

Also at the time of admission, laboratory studies

revealed an antistreptolysin-O titer of 531 IU/mL (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 remis-

sion 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 find-
ings, 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 pediat-

ric cases in the acute phase and during recovery, and

demonstrated the full reversibility of bilateral basal gan-

glia 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 semi-

quantitative 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 cor-

relate two sequential PET studies in an adult patient.

Striatal hypermetabolism in our case may have been either

direct consequence of an underlying pathogenesis or an
autoimmune-mediated inflammatory process leading to

enhanced corticosstriatal synaptic activity. In other autoim-

mune chortic disorders, such as primary antiphospholipid

syndrome and systemic lupus erythematosus, unilateral striatal

hypermobolism 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 natu-

ral precedent for direct immunoglobulin-mediated neuronal

activation, antibodies able to stimulate glutamate receptors

have been created in experimental models [8]. Further-

more, the absence of hypermetabolic basal ganglia findings
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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: FEW (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 disclose 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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