Neuropsychiatric disorder with basal ganglia lesions

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DESCRIPTION

A 15-year-old boy with normal birth and developmental history developed fever with a sore throat, which subsided within 5 days by over-the-counter medications. Fifteen days after the resolution of fever, he developed abrupt onset vocal and motor tics with choreiform movements of his left hand, hypersexuality and nocturnal enuresis in temporal association with group A beta-haemolytic streptococcus. Possibilities such as autoimmune encephalitis like N-methyl D-aspartate (NMDA) encephalitis, Anti phospholipid antibody (APLA) syndrome, neurosarcoidosis, Bechet syndrome, mitochondrial cytopathy, Wilson's disease, post-encephalitis sequelae, neoplastic lymphoma/glioma, and paediatric autoimmune neuropsychiatric disorder associated with streptococcal infections (PANDAS) were kept, and he was evaluated.

MRI of the brain was done (figure 1). Cerebrospinal fluid (CSF) analysis cellularity and biochemistry were normal, and the evaluation for viral, bacterial, mycobacterial and fungal encephalitis was unyielding. Serum copper studies and ceruloplasmin levels were within normal limits. The serum and CSF evaluation for autoantibodies against neuronal cell surface antigens were negative. The antinuclear antibody profile and paraneoplastic profiles were also negative. The anti-DNase B titres were elevated 682 (<200), while the antistreptolysin O (ASO) and antibasal ganglia antibodies were negative. Echocardiography was also normal. Whole mitochondrial exome sequencing and clinical exome sequencing for chorea were unremarkable. The patient's brain imaging was repeated after 6 months of initial symptoms (figure 2).

The patient had stereotypical motor movements and was using swear words and choreiform movements of the limbs in temporal association with a group A streptococcal infection, suggested by the positive anti-DNAase B titres. He fulfilled the diagnostic criteria for PANDAS.[1] It is important to note that elevated anti-DNase B titres occur in 80%, while the ASO titres are elevated only in 20%–50% of cases.[2]

Neuroimaging is usually normal in PANDAS. However, basal ganglia enlargement and hyperintensities can be seen in some cases.[3–5] The MRI brain showed bilateral striatal necrosis in this patient (figure 1). Such an imaging picture could be seen in mitochondrial cytopathies, amino-acidurias, Wilson's disease and parainfectious striatal necrosis. These disorders were ruled out by genetic, metabolic and antibody testing.

The disorder is self-limiting in most cases and can have a fluctuating course in relation to recurrent streptococcal infections. Symptomatic management with antipsychotics, benzodiazepines and valproate might be helpful. In addition, corticosteroids, intravenous immunoglobulin and plasma exchange have been tried in case reports. In a randomised controlled study, it was found that plasma exchange led to 48% and IVIg led to 41% improvement in clinical global impression score at 1 month, as compared with placebo, which did not lead to any change in scores.[6] Treatment with corticosteroids has been found to reduce symptoms by 3.5 weeks compared with those not treated who had a median duration of 11.4 weeks in observational studies in flares associated with recurrent streptococcal infections.[7] Our patient was started on risperidone, sodium valproate, clonazepam and prednisolone 40 mg. He was followed up after 2 weeks. There was a complete improvement in nocturnal enuresis, 80% in sleep, 60% in urinary frequency and 30% in motor tics and hypersexuality. However, there was no improvement in vocal tics and chorea. The

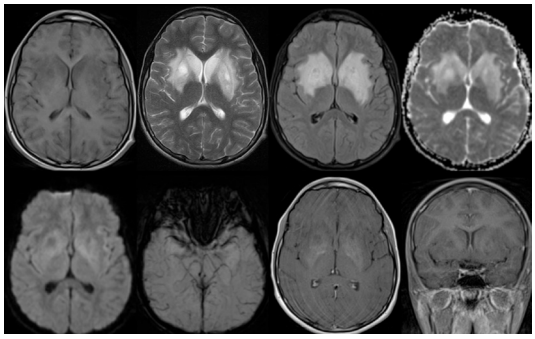

Figure 1 Bilateral symmetrical T2/FLAIR hyperintensities with swelling of bilateral basal ganglia, internal and external capsule, claustrum and extreme capsule with sparing of the insula; no diffusion restriction with minimal contrast enhancement.

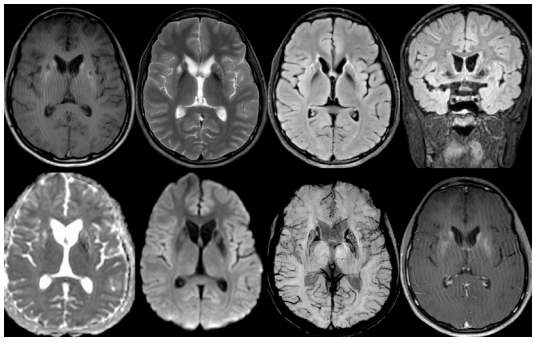

Figure 2 MRI brain 6 months later: bilateral striatal necrosis with resolution of T2/FLAIR hyperintensities of the above areas, bilateral striatal T1-hyperintensity without any enhancement. FLAIR, Fluid attenuation inversion recovery.

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Learning points

- Paediatric autoimmune neuropsychiatric disorder associated with streptococcal infection (PANDAS) is a cause of tics in children.
- Elevated anti-DNase B titres are more sensitive and occur in 80%, antistreptolysin O titres are elevated in 20%–50%, and antibasal ganglia antibodies in around 40%.
- Neuroimaging is usually normal in PANDAS. However, rarely, basal ganglia enlargement and T2 hyperintensities can be seen, as in this case.

patient's caregivers chose to observe for a few weeks and decide regarding further immunotherapy later.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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