but not *DMD*, had mental retardation if *IL1RAPL1* (interleukin-1 receptor accessory protein-like gene 1) was deleted.[4] The diagnosis of GKD–DMD is essential to foresee metabolic decompensations. Life-threatening episodes in childhood can be avoided by frequent carbohydrate meals and avoidance of excessive physical activity.[1-3]

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Idiopathic anti-NMDA-receptor encephalitis in a young Indian girl

Sir,

Anti–N-methyl-D-aspartate-receptor (NMDAR) encephalitis is a recently described paraneoplastic syndrome often associated with ovarian teratoma or idiopathic autoimmune encephalitis. It is associated with antibodies against NR1-NR2 hetromers of the NMDAR in serum and cerebrospinal fluid (CSF).[1] We describe the first case of idiopathic anti-NMDAR encephalitis from India.

A 13-year-old girl developed acute-onset behavioral abnormalities, gait disturbance and bruxism over a period of 1 month. An initial EEG and contrast magnetic resonance imaging (MRI) were normal. At admission, she was stuporous and mute. EEG showed frequent electrographic seizures of possible right-hemispheric onset. She was mechanically ventilated and required multiple anticonvulsants, including midazolam infusion for five days, before seizures could be controlled. CSF examination revealed 32 lymphocytes; protein, 52 mg/dL; and normal sugar. An extensive CSF encephalitic panel was negative. Twenty days after admission, she developed orofacial grimacing movements with oculogyric deviation and generalized chorea. Repeat contrast MRI of brain was normal. Subsequent multiple EEGs were normal. For another month, she continued to have episodic hypoventilation and cardiac dysautonomia. At this point of time, serum anti-NMDAR (N-methyl-D-aspartate-receptor) antibody testing was reported as positive (Prof. Angela Vincent, Weatherall Institute, Oxford). An 18F FDG PET-CT showed minimal-to-moderate FDG uptake in multiple lymph nodes, bone marrow and spleen, suggesting the possibility of a lymphoproliferative disorder, but no evidence of a teratoma [Figure 1]. There was hypometabolism in the left temporal and both occipital lobes [Figure 2]. Intense diffuse FDG uptake in both the lungs was attributed to nosocomial pneumonia. Multiple cervical lymph-node biopsies and bone marrow aspiration biopsies showed only reactive changes. These findings made us consider the diagnosis of idiopathic anti-NMDAR encephalitis. She was treated with two courses of IV methyl-prednisolone 1 g × 5 days, followed by IVIg 2 g/kg. Two weeks later, she became more responsive and was weaned off the ventilator. Five months later, she was ambulating independently and communicating with gestures.

Anti-NMDAR antibodies bind to the NR2B or NR2A

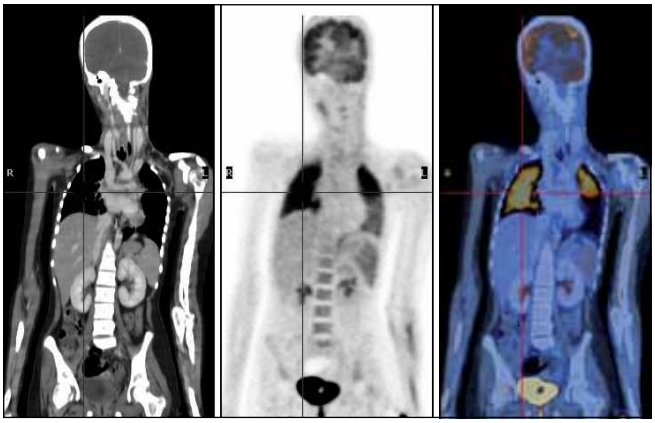

Figure 1: FDG PET-CT coronal images showing uptake in the lungs

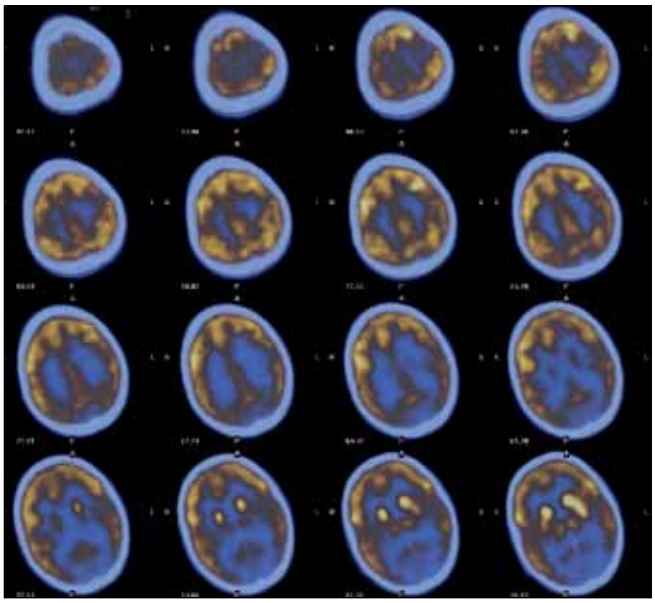

Figure 2: FDG PET-CT axial images of brain showing hypometabolism in the left temporal and bilateral occipital lobes

subunits of NMDAR. NR2B binds glutamate and is avidly expressed in the hippocampal and forebrain neurons of human beings.[2] These antibodies are thought to inhibit NMDARs in presynaptic GABAergic interneurons, resulting in reduced GABA release and disinhibition of postsynaptic glutamatergic transmission with excessive release of glutamate in the prefrontal/ subcortical structures. The pathogenic role of these antibodies is further strengthened by their disappearance during clinical improvement.

The typical clinical evolution of anti-NMDAR encephalitis includes five phases: phase I (prodromal phase)- 'viral-like' illness; phase II- acute psychosis and behavioral symptoms; phase III- intractable seizures, central hypoventilation and dysautonomia; phase IV- hyperkinetic phase with orofacial grimacing; and phase V- gradual recovery from the illness. MRI of the brain is either normal or shows nonspecific changes.[3] EEG shows only diffuse slowing. CSF shows features of inflammation with pleocytosis, increased protein and oligoclonal band.

ANMDARE was first described in a female in 1997 as ovarian teratoma–associated limbic encephalitis (OTLE). Besides, as a paraneoplastic syndrome, this disorder can be idiopathic in 30% to 40% of patients and has also been reported in males.[4] Rarely anti-NMDAR encephalitis has been associated with malignancies such as Hodgkin's lymphoma or testicular teratomas.[5,6] The treatment is removal of the underlying neoplasm, combined with immunotherapy, plasma exchange, intravenous immunoglobulin, and corticosteroids. Idiopathic anti-NMDAR encephalitis may show a poor response to treatment.

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Gradual onset of dyskinesia induced by mirtazapine

Sir,

Mirtazapine is a noradrenergic and specific serotonergic antidepressant (NaSSA) and is approved for the treatment of major depressive disorder and also has the potential to be of use in other psychiatric disorders.[1] Cases of acute onset dyskinesia,[2] dystonia,[3] and akathisia[4] have been reported with mirtazapine treatment. In this report, we present a patient who developed gradual onset of dyskinesia on mirtazapine.

A 76-year-old woman was diagnosed with depression and anxiety disorder in December 2006 and was put on clonazepam with a positive effect on anxiety symptoms; however she discontinued the medication. In September 2007, mirtazapine (15 mg/day) was added. On 26 October 2007, her general practitioner noted "flared" movements