Isolated Tongue Involvement—An Unusual Presentation of Wilson’s Disease

Sir,

The clinical picture of Wilson’s disease in children can be extremely varied, and includes all forms of acute and chronic liver disease, minimal to severe neurological disease, psychiatric problems, bony deformities, and hemolytic anemia and endocrine manifestations. The most common presentation of Wilson’s disease in children is liver involvement. We report a girl who presented with isolated tongue involvement as the sole manifestation of Wilson’s disease.

An 11-year-old girl presented with history of slurred speech and abnormal tongue posture for one month. She also had difficulty in swallowing. There was no past history of fever with joint pain or jaundice. She had been started on phenobarbitone and chlorpromazine for tongue symptoms.

On examination her vital signs were stable. There was no pallor, icterus or significant lymphadenopathy. Central nervous system examination revealed a conscious and oriented child with no cranial nerve palsy but the tongue was kept protruded out. She was able to withdraw the tongue back on command. Her motor, sensory and cerebellar systems were normal. Other systemic examination didn’t reveal any abnormality.

Hemogram revealed hemoglobin concentration of 12 gm%. The white blood cell count was 5900 cells/μL (Neutrophil 56%, Lymphocytes 30%, Band forms 7%, Eosinophils 4%) and Platelets count was 1,74,000/μL. The concentrations of electrolyte and creatinine in the serum were normal. Serum concentration of bilirubin (total) was 0.4 mg/dL (direct fraction 0.2 mg/dL) and of proteins was 7 gm% (albumin 4 gm%). Serum levels of AST, ALT and alkaline phosphatase were 43, 25 and 184 U/L. Her prothrombin and activated partial thromboplastin time were within normal limits. Her serum ceruloplasmin was 5 mg/dL, serum copper level was 48 μg%. Her 24-hour urinary copper was 166 mg in 1280 ml and after penicillamine challenge increased to 599 mg in 1550 ml. Slit lamp examination of eye revealed Kayser Fleischer ring.

Diagnosis of Wilson’s disease with neurological manifestation was made on the basis of presence of tongue dystonia, Kayser Fleischer ring in the eye, low ceruloplasmin and positive penicillamine challenge test. She was started on D-penicillamine 250 mg twice a day and pyridoxine 25 mg once a day. She was advised to avoid food with high copper content such as chocolate, nuts, legumes, mushrooms shellfish and liver. Her tongue dystonia disappeared but slurred speech persisted.

Neurological onset in Wilson’s disease has been record in children as young as 6 years and in adults as old as 52 years.[1] Neurological symptomatology is generally limited to motor system, presenting as manifestations of extrapyramidal or cerebellar dysfunction. [2] Extra-pyramidal symptoms can include facial grimaces, stereotypic gestures, dystonia, drooling, fixed grim, dysphagia and jaw or extremity contractures. Copper deposits in gray and white matter along with the basal ganglia. In our case, tongue dystonia presenting as sole manifestation, which is an unusual presentation of Wilson’s disease. Tongue involvement could also be due to basal ganglia involvement. In 2001, Singha Topaloglu presented a pediatric case with tremor of tongue and dysarthria as the only finding in Wilson’s disease.[3] In 1991, Liao and colleagues described a 15-year-old boy with involuntary tongue movement and speech disorder as an early manifestation of Wilson’s disease.[4] There are only a few case reports with tongue dysfunction as a sole manifestation of Wilson’s disease in literature and our case is one of them.

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References