Letter to Editor

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Pharmacology Section

An Early Sign of Wilson's Disease: Dysarthria

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Dear Editor,

We would like to present here, a case of Wilson's disease (WD), who was having a history of dysarthria since 3 months. A 10-year-old male child was admitted to Neurology Department of a tertiary care hospital in northeast India. As per the medical history of patient which was narrated by his mother, patient had been having episodes of high grade fever, loose motions and difficulty in speaking, for around 3 months. After consulting a private practitioner, fever and loose motions of the patient subsided, but slurred speech was persistent. Gradually, patient developed a low pitched voice, was unable to hold his footwear because of abnormal posturing of left leg toe and he had difficulty in chewing and swallowing.

On examination, his vital signs were normal, except icterus and pallor. Ceruloplasmin is a serum glycoprotein which carries majority of copper in the blood and the patients with WD have low levels of ceruloplasmin [1]. In contrast, in our case, serum ceruloplasmin was within normal range, i.e. 324mg/l (normal 200-350mg/l), whereas serum copper was 46µg/dl (normal 70-140µg/dl). The haemoglobin concentration was 16gm%, white blood cell count was 9.1×103/mm^3 and platelet count was 106×103/mm^3. The serum concentrations of electrolytes were normal, creatinine was 0.36mg% and total bilirubin was 0.3mg% (direct fraction 0.2mg). Serum levels of AST, ALT and alkaline phosphatase were 88, 65 and 242U/L respectively. The 24 hour urine copper level was 835.48µg/day (normal 32-64µg/day). The diagnosis was made on the basis of urine copper levels, Kayser-Fleischer ring and face of giant panda on T2-weighted images seen on Magnetic Resonance Imaging. The penicillamine test was not performed in this case.

At the hospital, tablet distamine (penicillamine 250mg) and tablet zincolac (folic acid 1.5mg, pyridoxine 3mg, riboflavin 10mg, thiamine 10mg, calcium pantithenate 5mg, lacto bacillus sporegens -60 million spores, niacinamide 45mg and zince sulphate 45mg) were prescribed to the patient and he was then discharged after one week. Thus, we can depict that tablet distamine was prescribed as a chelator to form penicillamine-copper complexes, while tablet zincolac was prescribed to block copper absorption from the intestinal track. However, after a few days, patient was again admitted with the complaint of aggravated abnormal behaviour and dystonia which involved tongue, right upper limb, left lower limb and right foot. The clinical features indicated potential intolerance to penicillamine and physician advised the patient's mother to withdraw the tablet distamine. Tablet trientine dihydrochloride (triethylenetetramine 1000mg) was prescribed instead.

In WD, copper accumulates in gray and white matter along with the basal ganglia, resulting in improper modulation of the basal ganglia by the subthalamic nucleus [2]. The mixed dysarthria with hypotonia and dystonia is common, because copper damages the extrapyramidal pathways as well as various components of motor system [3]. Initially, patient had difficulty in pronouncing lingual and he gradually developed a slower speech rate, hypophonia, variation in loudness, abnormal stress patterns on words, stuttering and palilalia. Thus, dysarthria in patients can be considered as an effective monitor of progress in the treatment of this disease [4]. This case indicated that dysarthria should not be overlooked in patients and that its cause should be figured out, for early detection of disease and to begin appropriate treatment.

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