Movement Disorders

WILSON`S DISEASE WITH NEUROPSYCHIATRIC MANIFESTATIONS IN CHILDREN: REFLEXION ABOUT A CASE

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Introduction:
Wilson`s disease, an autosomal recessive genetic disorder, causes tissue accumulation of copper initially in the liver and then in the central nervous system responsible for neurological complications. The diagnosis is clinical, biological and molecular. Various chelating treatments are available to decrease the spontaneous morbidity and mortality of this condition.
Material and method:
We report the clinical observation of an 11-year-old child followed at the child psychiatry department of Ar-razi hospital in salé for a wilson`s disease with initial neuro-psychiatric manifestation.
Discussion:
Neuropsychiatric manifestations are present in 35% of Wilson`s disease cases with dystonia, extrapyramidal syndrome, dysarthria, dysphagia or psychiatric manifestations. The delay in diagnosis remains long, responsible for the severity of the disorders and the poor response to treatment. The first-line treatment appears to be chelation therapy with triethylene tetramine (Trientine®), which is less often responsible for side effects and initial worsening of symptoms than D-penicillamine. Zinc is the first-line treatment in asymptomatic forms and as maintenance therapy. Finally, liver transplantation may allow clinical improvement.
Conclusion:
The diagnosis of Wilson`s disease with neurological expression is difficult. It must be evoked in front of various neurological and psychiatric signs. Delay in diagnosis is the main unfavorable prognostic factor.