Hallervorden-Spatz Disease

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Hallervorden and Spatz in 1922 described an autosomal recessive disorder in which there is progressive CNS degeneration with predominant changes in the basal ganglia. To the best of our knowledge, this disease has not been reported from India.

Case Report

A nine-year-old boy, 4th born to a non-consanguineous parents presented with history of involuntary movements with dystonic posture for one and half years, and progressive deterioration in locomotion, speech, swallowing, sphincter control alongwith impairment of vision for four months. Antenatal, natal and post natal periods were uneventful, child had regression of attained milestones. An elder sister had similar illness and died at the age of nine years.

Examination revealed a malnourished child with normal head circumference and severe choreo-athetotic movements mainly involving the upper extremities, alongwith dystonic arching of the trunk and drooling of saliva *(Fig. 1)*. Deep tendon reflexes were hyporeflexic and superficial reflexes were absent with extensor plantar response. Fundus examination revealed bony pigmentary spicules alongwith attenuated Vessels and pale disc, suggestive of retinal pigmentary dystrophy without any evidence of cherry red spot. On slit lamp examination, there was no evidence of Kayser-Fleischer rings.

Peripheral smear showed acanthocytosis *(Fig. 2)*. Serum copper, ceruloplasmin, iron and electrolytes, lipoproteins and cholesterol were normal. Skull X-ray and CSF were within normal limits. EEC showed generalized slowing without any spike wave discharge. Bone marrow examination was normal.

Discussion

A clinical picture of progressive neurological degeneration involving the basal ganglia(l), associated fundus and peripheral smear findings(2) without any evidence of Wilson's disease with similar illness in another sibling suggestive of autosomal recessive mode of inheritance, aroused the clinical suspicion of Hallervorden-Spatz disease.

Pathologically there are characteristic abnormalities in the basal ganglia consisting of pigment deposition of mainly iron in globus pallidus and substantia nigra alongwith focal axonal swelling(l). There is no associated systemic disorder of iron metabolism. Studies on labelled iron kinetics have shown a selective uptake of iron in the region of basal ganglia(2). The co-existence of Hallervorden-Spatz disease with

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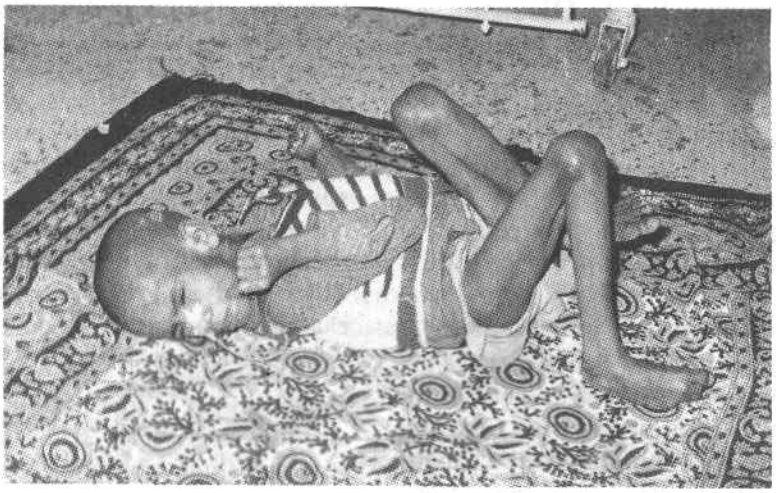

Fig. 1. Photography of the malnourished child with abnormal posture.

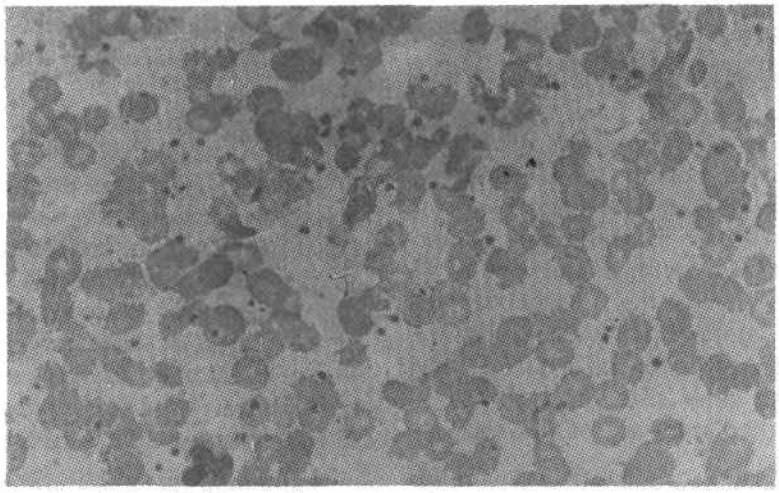

Fig. 2. Microphotograph of blood smear shoxving acanthocytes with characteristic spurs.

acanthocytosis has been previously described in one family(3). Sea blue histiocytes have been described in bone marrow aspiration, but they are nonspecific and seen in various other neurological disorders. Magnetic resonance imaging scan shows a characteristic pattern of increased density in globus pallidus surrounded by low density on T_2 weighted images known as the "eye of the tiger" sign(4).

There is no definitive treatment for this condition, though chelating agents, levodopa and other antiparkinsonian medications have been tried without much success. There is progressive neu-

rological deterioration and most of the children die by 10 years after the onset of neurological deficit.

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Oral Acyclovir in Treatment of Suspected Herpes Simplex Encephalitis

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Herpes simplex encephalitis (HSE) is a relatively frequent cause of sporadic acute encephalitis associated with progressive neurologic signs(l). It is the most common cause of fatal sporadic encephalitis and it accounts for 33-57% of cases of encephalitis with focal localizing signs. In survivors, it frequently produces severe sequelae(2). Intravenous acyclovir is currently the drug of choice for HSE and it substantially reduces the mortality and morbidity(2). We could not come across any reports of trials of oral acyclovir in the treatment of HSE.

We report here, a case of suspected herpes simplex encephalitis, which responded dramatically to oral acyclovir therapy. The cost of 10 days treatment of a 25 Kg child with intravenous therapy drug is Rs. 21,000/-, while it is only Rs. 750 with oral therapy.

Case Report

A 2 year 10 months old girl was admitted with complaints of sudden onset of altered sensorium, fever and left sided recurrent focal seizures for the past 7 hours. There were no overt manifestations of herpes simplex infection in the patient or in her contacts and no local outbreaks of similar illness. There was no history of head injury, ear discharge, local infection, and dog bite.

On admission, the patient was

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