Letters to the Editor

Hallervorden Spatz Disease

The case report on Hallervorden Spatz disease (HSD)(1) aroused interest, particularly because we are currently managing a child with this disease in our Pediatric Neurology Clinic. Although the authors have rightly suspected the condition, they have not presented parameters on the basis of which the diagnosis was made. Absence of criteria for Wilson's disease and presence of family history of similar illness, does not automatically make this a case of HSD. There are other causes of progressive dystonia which may also be familial. Acanthocytosis is not a feature of HSD; in fact only 2 out of 64 cases of HSD reported earlier showed acanthocytosis(2). Familial acanthocytosis, however, is an important hereditary neurodegenerative disease associated with progressive dystonia.

Inspite of various parameters having been studied, there are, as yet, no specific biochemical or other markers of the disease *in vivo* and the diagnosis has generally been made post-mortem. Increased uptake of iron in basal ganglia on radioactive iron studies has been suggested as a helpful test(3) but such studies are cumbersome. CT scan provides non-specific findings; ventricular dilatation and increased density in basal ganglia are reported (4). The only modality by which the diagnosis can be made with reasonable certainty during life is the MRI scan which shows low signal intensity in the globus pallidus on T2 weighted SE and RE images and in some cases the so called 'tiger-eye' appearance(5). As the authors have not mentioned any findings of CT or MRI scan, making this specific diagnosis is not justified.

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Reply

We considered all heredo-degenerative disorders of the basal ganglia in our case before diagnosing Hallervorden-Spatz disease. In dystonia musculorum deforman, intellect remains normal(l). Our case was having significant dementia.

Fahr's disease is characterized by choreo athetotic movements, microcephaly and basal ganglia calcification(2). Acanthocytosis and retinitis pigmentosa are rare and prominent findings in our case, association of which has not been described with any of the heredo-degenerative disorders of the basal ganglia except occasionally with Hallervorden disease(2).

Hereditary acanthocytosis was excluded as it is associated with extreme proprioceptive sensory loss, ataxia, steatorrhea and low serum cholesterol(3). Wilson's disease was also excluded in the presence of normal serum copper and ceruloplasmin levels.

Demonstration of iron retention in the basal ganglia is a specific though cumbersome test. MRI findings as mentioned by Dr. Singhi are characteristic and have been used recently for the diagnosis. The diagnosis must, however, be suspected clinically and patients referred for evaluation.

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NOTES AND NEWS

UPDATE ON COMMON PROBLEMS IN PEDIATRIC PRACTICE

The IAP Baroda Branch is organizing this event on 21 April, 1996 at Hotel Surya Palace, Vadodara. The registration fee for delegates is Rs. 150/- and for postgraduate students is Rs. 100/- upto 13th April, 1996. Spot registration is Rs. 200/- and Rs. 150/-, respectively. Cheques and DD must be drawn in favor of Indian Academy of Pediatrics. Baroda Branch and sent to: Dr. Sangita Trivedi, Organizing Secretary, 2, Amar Complex, Chanakyapuri Road, Sama, Baroda 390 008. Phone: (C) 482694 (R) 482616.