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Wichman Syndrome Simulating Posterior Fossa Mass in CT Scan

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Congential cerebeller hypoplasia as an autosomal recessively inherited disorder has been reported by Wichman *et al.*(1). This syndrome comprises of gait ataxia, hypotonia, mental retardation, cerebellar hypoplasia, ventricular dilatation and vermis anomalies. Three sibling pairs with abnormalities in CT Scan ranging from prominant valleculla to enlarged cisterna magna with hypoplasia of cerebellar hemispheres and vermis have been reported, the pedigrees were consistent with autosomal recessive inheritance(1). Seven more children with congenitally small cerebella have also been reported as isolated cases described to be sporadic, nonprogressive malformations of the brain(2).

We present here an isolated case undetected till the age of 9 years with similar features. However, the point to be kept in mind is the glaring disparity between the CT Scan and MRI findings.

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Case Report

A 9-year-old female, presented with staggering gait drifting to the left from early childhood. There was history of occasional episodes of vomiting, headache and vertigo since 6 years of age, each episode lasting for 2-3 days. Two recent attacks were preceded by fever, malaise, watering and redness of eyes. There was no history of seizures, altered sensorium, visual disturbances of deafness. Speech, swallowing and chewing were unaffected. Scholastic performance was

poor throughout. Antenatal, intranatal, and postnatal history was uneventful. There was no similar family history or consanguinity between parents.

Examination revealed an IQ of 80 (Binet test), mild facial asymmetry (left palpebral fissure smaller than the right), normal shape of skull, no cranial bruit and head circumference of 50 cm. Systemic examination was within normal limits. CNS examination showed normal speech, hearing and vision; and fundus was within normal limits. Pupils were normal in size and reaction. There was no nystagmus but saccadic eye movements were slow to left gaze. Other cranial nerves were within normal limits. Hypotonia was present in left upper and lower limbs within power grade 4+/5. All reflexes were normal with flexor planter responses. There was no sensory loss. Cerebellar signs in the form of difficulty in tandem walking and left sided dysdiadochokinesia were present.

Investigations revealed: Hb 11.9 g/dl, ESR 100 mm/h, TLC 5.9 × $10^3/\mu l$ with a Mantoux test of 23 × 22 mm (72 hrs later). Chest *X*-ray showed calcified perihilar lymphnodes. CSF studies revealed 8 cells/mm^3 (all lymphocytes), protein 70 mg/dl, sugar 63 mg/dl and sterile culture (including AFB).

CT Scan head (contrast enhanced) showed an enhancing lesion in the posterior fossa (left) with irregular edges coming upto the inner calvarium. Anteriorly, the lesion extended along left tentorial margin (*Fig. 1*).

Magnetic response imaging of brain was done on a 1.5 tesla superconducting unit (Megnatom, Siemens, Germany). T_1 (TR/TE = 500 msec/15 msec), T_2 (2800/80) and proton density (2800/20) weighted images were obtained in axial, sagittal and coronal planes. This revealed partial agenesis of left cerebellar hemisphere and lower vermis angulation of brain stem (*Figs. 2a & b*). There was assymetric ventricular dilatation with aqueductal stenosis. Post contrast study with Gadolineum-DTPA revealed enhancement of the left transverse sinus, similar to

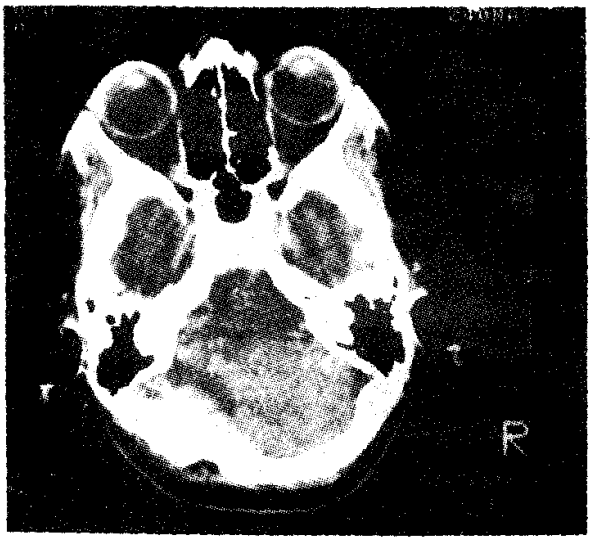

Fig.1. Contrast enhanced axial CT Scan head through the posterior mass on the left side in relation to left cerebellum adjacent to the left occipital bone.

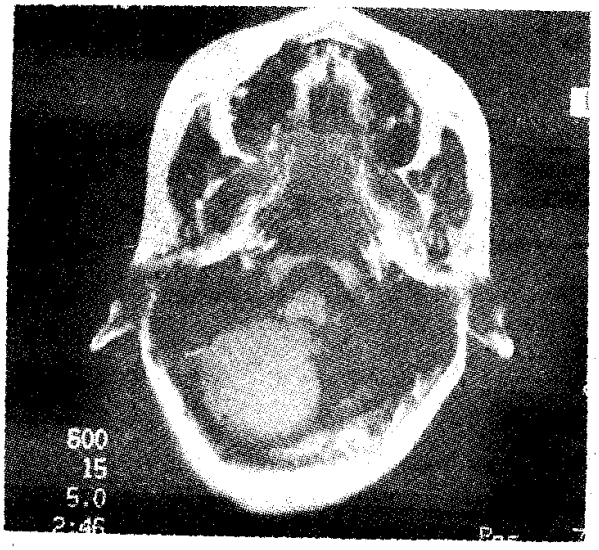

Fig.2a. The axial T_1 weighted MRI image shows absence of left cerebellar hemisphere.

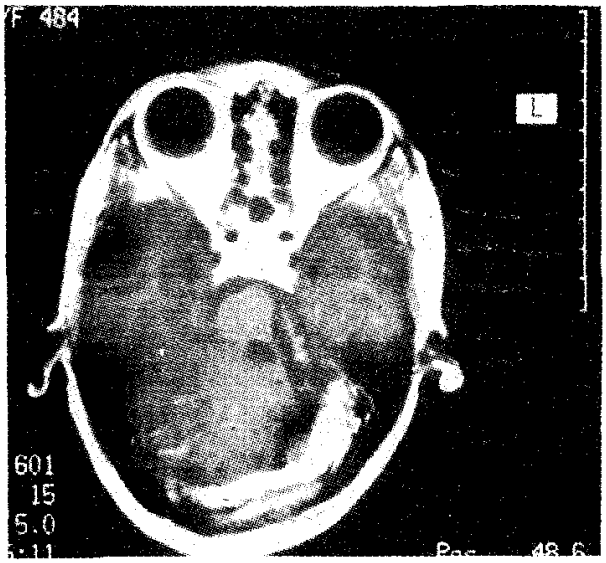

Fig. 2b. Post contrast Gd-DTPA axial T1 weighted image shows dilated left transverse sinus with atrophy and angulation of pons.

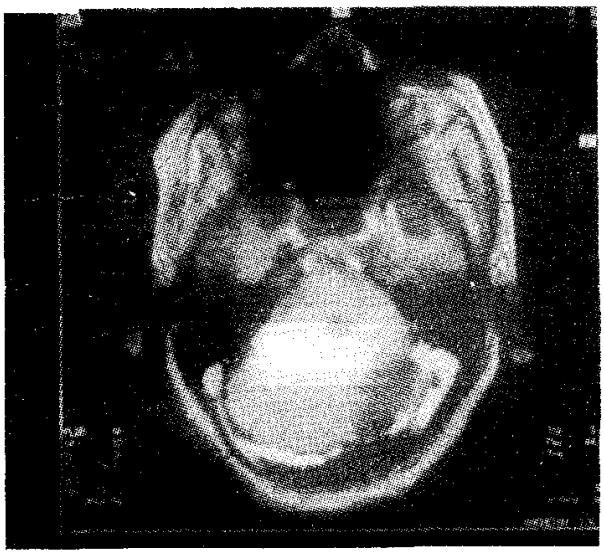

Fig. 2c. Flash image shows the left transverse sinus seen as bright signal (Arrow).

what was seen in post contrast CT. Flash images in axial plane revealed (22/10/40°) encoding of blood in the transverse sinus (*Fig. 2c*).

Discussion

The patient reported presently demonstrated non-progressive cerebellar dysfunction with mental deficiency (IQ 80). There have been some reports of cerebellar hypoplasia(2,4) but all were associated with profound mental deficiency. An autosomal recessive disorder with cerebellar hypoplasia has been reported in 3 pairs of sibs(1). The patient in the present report showed subtle motor clumsiness with subnormal intelligence. This seems to be more consistent with the autosomal recessive pattern which may show marked clinical heterogeneity.

Neuropathologically, similar cases have shown severe degeneration of granular cells with mild to moderate degeneration of purkinje cells. The centre of the cerebellar white matter had mild pallor or was usually not affected(2). Some reports also document loss of neurons in the cerebellar hemispheres. The mechanism of these abnormalities is not known but such have also been reported in experimental animal models(5).

The other interesting aspect is simulation of the case with a left posterior fossa enhancing mass in CT Scan head. However, cranial MRI detected the malformation as described; the apparent enhancing mass was dilated left transverse sinus (compensatory to volume loss due to ipsilateral cerebellar hypoplasia).

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Ultrasonographic Study in Meningitis

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Cranial ultrasound has been shown to give useful information of intracranial pathology in various meningitides in infants and young children(1-9). We studied cases of acute bacterial and tuberculous meningitis ultrasonographically to find out if this technique could be useful in: (*a*) suggesting the cause, *i.e.*, acute bacterial or tuberculous; (*b*) monitoring the course; and (*c*) indicating the outcome of these diseases.

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Material and Methods

Thirty five consecutive children with meningitis, 20 acute bacterial (ABM) and 15 tuberculous (TBM) with open anterior fontanallae (AF) admitted into the pediatric wards were studied prospectively by cranial ultrasonography (CR-USG) soon after the initial diagnosis. ABM was diagnosed by: (*a*) suggestive clinical picture; (*b*) routine CSF examination showing polymorphonuclear pleocytosis, protein >40 mg/dl and sugar <40 mg/dl; and (*c*) positive CSF or blood culture or positive latex agglutination test (this however was not considered a must). TBM was diagnosed by: (*a*) suggestive clinical picture; (*b*) routine CSF examination showing lymphocytic pleocytosis, protein >40 mg/dl and sugar <40 mg/dl; and (*c*) finding of at least one of the following evidence of tuberculosis–positive tuberculin test, positive *X*-ray chest, positive contact survey and recovery of AFB from gastric aspirate or CSF.

CR-USG was done by Siemen Sonolyne SL II Real-Time Grey Scale Sector Scanner with a 5 MHz transducer using standard technique. AF was used as the acoustic window. The scans were performed in coronal, sagittal and parasagittal planes. The time gain compensation controls were set to obtain adequate penetration to pick up even the far field parenchymal echoes. During the scans,