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Case Report-1

NPH Presenting As Parkinsonism And Early Optic Atrophy

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Abstract :

An old hypertensive patient presented with progressive slowness of movement and difficulty in walking for 6 months. He also complained of the gradual memory loss with blurring of vision (left eye more than right) and incontinence of urine. Patient was confused with slow, indistinct speech and lead-pipe and cogwheel rigidity in both upper and lower limbs. Mild weakness in left side of body with left plantar extensor. Primitive reflexes were present. CT scan of brain showed multiple lacunar infarcts with grossly dilated 3rd, 4th and lateral ventricle with rounding of frontal horns and minimal dilatation of sulci. Finally he was diagnosed as parkinsonism secondary to NPH (Normal pressure hydrocephalus). Although extrapyramidal feature in NPH are not uncommon, presentation with parkinson syndrome as a predominant feature is rare. Failure of the patient with parkinsonism to respond to levodopa therapy, should alert one about the possibility of NPH.

Introduction :

There is universal acceptance of Hakim and Adams[1] original description of the classical feature of normal pressure hydrocephalus; The trait of gait disturbance, dementia and urinary incontinence. There is wide variation in the clinical manifestations of NPH, particularly in regard to the disturbance of gait. This is most often as spastic ataxia[2], although less commonly, parkinsonian features can occur[3]. Rarely patients may present atypically with signs, indistinguishable from Parkinson syndrome[45]. We report a case of NPH which present predominantly as parkinsonism, improved bestly by the repeated CSF withdrawal. Clinical motor signs of NPH subjects extend beyond gait difficulty which include extrapyramidal manifestation of bradykinesia, akinesia, rigidity and propensity to perform more poorly when external cues to move are absent of this parkinsonian and Huntingtonian movements and these signs may reverse following therapeutic CSF drainage[6 7]. Neuro-opthalmologic signs include optic atrophy, visual field defect,abnormal pupils, papilloedema and visual agnosia[8]. One interesting feature is in NPH patient; when increased ICP intermittently at night, then altered CSF dynamic thereby motor and neuro-opthalmological feature can develop. There are several anecdotal reports of NPH causing parkinsonian motor behaviour[9].

Case Report : A fifty two years old, hypertensive , right handed person presented on 9th December 2009 with the history of progressive slowness of movement and difficulty in walking for six months. He also complaints loss of vision, more marked on left side with loss of memory and he was not able to do executive work. Five days prior to admission, he felt difficulty in walking with very short stepped gait with repeated fall. Acuity of vision markedly reduced in left eye and decreased colour perception. He also complaints the mild left sided weakness that developed 10 days prior to presentation. There was tremor in legs during standing and mild disequilibrium also present. He did not give any history of the fever, double vision or opthalmoparesis, head injury or difficulty in swallowing. There was not prior history of tuberculosis or no significant headache with SAH, convulsion or coma, no family history of the neurological disease. Personal history revealed no history of alcohol, any illicit drug, kobiraji drugs or history of exposure. On examination , he looked ill and masked face and stooped posture. Pulse was 88/min, BP was 150/95 mmHg, temperature was normal and other general examination parameters were normal. On systemic examination, all the systems seemed to be normal except nervous system which revealed, he was confused and reduced executive activity, speech was very soft, slow and indistinct. All other cranial nerves were intact except second and eighth cranial nerve which showed marked reduction of

visual acuity in left eye with disturbance of colour vision. On fundoscopy – bilateral optic atrophy which were more marked on left side with distinct border. Both pupil were mildly dilated and light reflex was sluggish. Right sided Weber test revealed sensorineural deafness. Muscle power reduced in left side, grade 2 in both upper and lower limb. His muscle tone showed lead pipe type rigidity in elbow and knee and cogwheel type in wrists. DTR in right side revealed normal and decreased in left side in both upper and lower limbs with left planter reflex extensor. There was no abnormalities in in-coordination or cerebeller examination. Position sense was impaired in both lower limbs and Romberg test showed no definite abnormality. On mini-mental state examination-MMSE was 19 with positive primitive reflexes. Patient didn't show any sign of meningeal irritation. His gait was short stepped, festinating type. Relevant investigation showed TC 11000, N-40%, Hb-8.1 gm%, ESR-50 mm in first hour, PBF-Eosinophilic Leucocytosis, RBS-116 mg/dl, Blood Urea-54 mg/dl, Serum Creatinine-1.8 mg/dl, Sodium-139.2 meq/L, K-4.15 meq/L. X-Ray chest P/A view, LFT, Urine RME, Lipid profile, VDRL, ELISA Test for tuberculosis, Vit-B12 assay, S TSH, TPHA and X-ray Lumbo-sacral spine showed no abnormalities. His ultrasonography of whole abdomen showed only enlarged prostate. CSF Cytology revealed total count 5/mm^3, Polymorph-0%, Lymphocyte-100%, RBC-8-12 HPF. In biochemical test protein 342mg%, Sugar 56mg%, CSF VDRL was non-reactive. CT scan of the brain showed dilatation of all ventricles with rounding of frontal horn with minimal atrophy of sulci. There was also multiple lacunar infarcts. We immedicately started Levodopa, Revastigmine, Amantadine and Anti-Hypertensive but one week later, features of parkinsonism did not improve. Then we were suspecting the case of NPH and repeated CSF withdrawn improve his gait and dementia with features of parkinsonism and finally diagnosed as parkinsonism secondary to NPH and was discharged with advise of subsequent follow up.

Discussion :

Normal pressure Hydrocephalus is a potentially reversible cause of dementia which is often amenable to surgical therapy[10]. Normal pressure hydrocephalus is an inaccurate term because long term monitoring of the patients with this syndrome have shown intermittently elevated pressure specially during the night[11]. The clinical feature of NPH are due to selective involvement of the periventricular region of brain[12], consequent upon progressive ventricular enlargement. Sypert et al [4] suggested that parkinsonian symptom in NPH are the result of mechanical distortion of the basal ganglia and consequent vascular insuffiency in the nigrostriatal system and in the same way , local distortion of the basal ganglia and midbrain by tumour can produce parkinsonian symtoms that disappears after successful decompression[13]. Patient may be misdiagnosed as having Parkinson disease because the gait disorder is similar in the two syndrome, suggesting that the origin of the problem in the hydrocephalic patient lies in the basal ganglion. Because many of this patient may also have hypertension and some have small or large strokes, such patient may have other neurological finding, including spasticity and hyperreflexia with babinski signs[10]. Our patient initially presented with gait disorder and slowness of movement as in parkinsonism. Chronic communicating hydrocephalus is now recognized as one of the causes for reversible dementia , with an occurrence of 1-10% among patient with diagnosis of dementia[14]. Our patient having significant dementia and reduced executive function. Neuro-opthalmologic findings in NPH have included optic atrophy, visual field defect, abnormal pupils, papilloedema, visual agnosia and subtle changes in disks (fullness of the veins with loss of a spontaneous pulses or early optic atrophy) should be searched for in suspicious cases[8]. CT and MRI of brain was aided in distinguishing Parkinson's disease, lacunars state and NPH, although NPH may occurring coexisted this disease[10].Our patient presented with bilateral optic atrophy with multiple lacunar infarcts. NPH may also follows SAH, head trauma, resolved acute meningitis or chronic meningitis (tubercular, syphilitic or other), pagets disease, mucopolysacharrides and achondroplasia[15]. Above this condition were excluded by the proper clinical search and relevant investigations. On CSF examination, our patient showed normal cytology and sugar but highly raised protein. The pathogenesis of the white matter changes in NPH and subcortical atherosclerotic encephalopathy(SAE) is different and ischemic white matter changes can be a part of the NPH state and the patient of NPH with concomittent cerebrovascular aetiology had higher sulphatide protein concentration in CSF and poorer outcome after shunt surgery than patients with other aetiology[14]. A slight plasma like protein pattern in CSF indicating a blood brain barrier dysfunction was seen in 38% of the patients before operation[16]. It was not considered that the causal mechanism of NPH was due to the high protein levels in the CSF. A patient with parkinsonian syndrome caused by NPH, if the CSFprotein levels high, the possibility of spinal cord tumour should be considered[17]. We advised patient MRI of

dorsolumber spine but patient failed to provide this. Patient was treated by the levodopa and repeated CSF drainage and his gait, slowness of movements and cognition were improved properly and discharged and refered for the shunt drainage , though concomitant lacunar infarct has lower rate of improvement after shunt drainage[10].

Conclusion :

The underlying motor pathophygiology of NPH signs extend beyond gait and bladder dysfunction with prevalence of extrapyramidal signs, this include even complex parkinsonian signs of reliance upon external Clues for improved motor performance. So it is concluded that we should suspect the parkinsonism may be due to NPH. Early detection and shunt treatment will cure the patient.

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