Dandy-Walker Syndrome with Epilepsy and Psychosis: An Atypical Presentation

S.C. Pradhan, Jnanamay Das & Vinod Kumar Sinha

Abstract

A case of "Dandy-Walker Syndrome" with secondary Generalised tonic clonic seizures and post ictal psychosis is reported in a 33 year old man. The ventriculo-peritoneal shunt procedure, carried on 3 years back has failed to bring down the seizure frequency. Electrophysiological and neuropsychological investigation suggest impairment in left temporal lobe.

Key Words: Dandy-Walker Syndrome, epilepsy, post-ictal psychosis

The Dandy Walker Syndrome (DWS) is either a developmental disorder or it is the residence of prior viral inflammatory disease contracted in utero or in early life. It is characterised by three main elements - posterior fossa cyst, the absence of the cerebellar vermis, and alteration in CSF dynamics. Though controversial the syndrome was reported to be described first by Virchow in 1863. Benda (1954) used the term 'Dandy Walker Syndrome', acknowledging the contributions of Dandy for clinicopathological descriptions and Walker for suggesting a possible surgical cure of the condition. Diagnosis is done by the clinical triad of cystic dilatation of the fourth ventricle, dysgenesis of the cerebellum, and CSF retention in the hindbrain.

DWS is considered to be a condition of multifactorial origin (Gardner, 1975). The chief etiological factors are hypothesized to be mutations, chromosomal aberration, mitotic interferences and altered nucleic acid synthesis. As pathogenesis of DWS usually starts in the embryonic period, more than 80% of the total cases are diagnosed within the initial year of postnatal life. Occasionally, the Dandy Walker malformation is seen in adults (Epstein et al., 1987) and a case has also been described at the age of 72 who did not have neurological symptoms until the age of 69 (Brown 1977). The earliest possible diagnosis is essential for optimal management and even survival. Murray (1983) and Depp et al. (1983) discussed the diagnosis of the Dandy-Walker syndrome in utero under ultrasonic guidance in a 30 week old fetus using ventriculo-amniotic shunt. At present the therapeutic modality of choice in DWS is ventriculo-peritoneal shunting. Mortality rate is between 25-45% (Epstein et al., 1987). Morbidity problems arise mainly from the complications with ventricular shunting.

The most obvious clinical feature in the Dandy Walker Syndrome is macrocrania followed by anterior fontanelle bulging, soft or sunken fontanelle and bossing of the forehead or occiput in some individuals. In view of the lack of development of the cerebellum, it is noteworthy that only a few patients (<15%) have signs indicative of cerebellar dysfunction, such as ataxia or nystagmus. Cranial nerve signs for example papilloedema and optic atrophy, III or IV nerve paresis are even rarer, whereas long tract signs are more common (Epstein et al., 1987). Symptoms such as irritability, vomiting.
headache, lethargy and seizures are relatively infrequent in their occurrence (approx. 10-20%). But there are no case report of DWS with psychosis, to our knowledge. Here, we present a case.

**CASE REPORT**

Mr. RD, a 33 year, old, married, Hindu, male from a rural background was admitted in Central Institute of Psychiatry, Ranchi with chief complaint of repeated short lasting unconsciousness with jerky movement of body for last 18 years and behavioural problems for last 3 years.

During the attack, the patient would become unconscious suddenly, would fall, body would become rigid first, and after 5-10 seconds, twitching of right angle of mouth would start, followed by jerky movement of the whole body for about a minute. He would gain his consciousness after about 5-10 minutes but remain confused for about 15-20 minutes. Followed by this he would sleep for 3-4 hours. During the attack there was history of frothing from mouth, superficial injury over body due to fall and incontinence of bladder only. There was no history of any prodrome and aura. Initially the attack used to occur 1-2 times in every 6 months, later the frequency increased to 1-3 times in every 2 months inspite of continued treatment with diphenyl hydantoin (DPH) in adequate doses.

After an operation was carried out by neurosurgeons in February 1991, the patient was put on diphenyl hydantoin (DPH) 300 mg and phenobarbitone 120 mg orally in daily divided doses. Though the patient remained symptom free for 6 months, after that he had jerky movement of whole right upper limb which used to start in the tip of index finger. Frequency of such jerky movements was 1-2 every month. Sometimes he would hear whistling, ringing of bell, even human voices for a few seconds associated with vertigo and pain abdomen. Most of the times these phenomena would invariably be followed by unconsciousness with jerky movements of whole body.

Abnormal behaviour was noticed for the first time about 3 years ago during post ictal phase. It was characterised by irritability, strong suspiciousness towards family members, using abusive languages, decreased sleep, decreased personal care, becoming undressed publicly, irrelevant talk and becoming assaultive at times. This phase used to persist for few days to few weeks following generalised tonic clonic seizure and would stop spontaneously. No change was noticed in appetite, bladder and bowel habit. There was no history of headache, nausea, vomiting, visual disturbances, lethargy, malaise and weakness.

About 20 years ago, he had a history of head injury after being trampled by an ox, which resulted in unconsciousness. He was taken to a local doctor, 5% Dextrose was given I.V. and he gained consciousness after 2 hours. Though two stitches had to be applied on the left parietal region of the skull, there was no evidence of fracture in the skull bones as corroborated by X-ray of the skull taken at that time. The patient had an uneventful normal delivery with normal developmental milestones and there was no history of neuropsychiatric illness in the family.

On general examination, he had mild occipital prominence of head. There was no protrusion of eye ball and prominent veins over skull. 'cracked-pot sign' was absent. No abnormality was detected in cardiovascular system, respiratory system & gastro-intestinal system. Nervous system abnormality included dysarthria and sensory neural deafness of left ear. In fundoscopy, no abnormality was detected, there was no evidence of papilloedema & optic atrophy, gaze paralysis was absent and there was no other abnormality detected in the cranial nerves. In motor system no loss of power and no abnormality in tone had been found. Wasting & abnormal movements were absent. No abnormality was detected in the sensory system too. Among the release reflexes, only palmo-mental reflexes were present bilaterally. All deep tendon reflexes could be elicited, were symmetrical on both sides and not exaggerated. Superficial reflexes were elicitable and plantars were down going
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on both sides. No abnormality was noted in gait, and the ventriculo-peritoneal shunt was intact.

His EEG has been done for the first time by neurologist outside at the onset of the seizures and for the second time after admission in our institute as a routine investigation in seizure disorders. Both the times EEG showed spike and sharp wave discharges from left temporal region becoming generalised. His I.Q. was found to be 78 which falls under borderline category. Luria-Nebraska neuropsychological battery (LNNB) revealed definite impairment in his left temporal lobe functions. The C.T. scan of head showed marked hydrocephalus involving dilatation of all the ventricles of brain. There was a growth resembling a cyst in the posterior fossa (fourth ventricle) which was interpreted as a Dandy-Walker cyst. Patient was treated by carbamazepine tablets 800 mg/day. His psychosis gradually improved and during discharge he did not have any active psychopathology. Seizures also remained controlled.

DISCUSSION

The patient had long continued generalised seizure which started after an event of head injury. Seizure is expected in a compromised brain and also after head injury, where as both the conditions are present together making brain more vulnerable for seizure. It may also explain the poor response to antiepileptic drugs. There was also simple partial seizure and complex partial seizure from the very beginning, but their manifestations were buried under generalized seizure. Thus, after the ventricular shunt operation when the frequency of generalized seizure decreased, partial seizure manifested.

Mild occipital prominence of head can be explained by the presence of cystic growth in posterior fossa of brain. Inspite of this there was no ataxia, nystagmus and other cerebellar signs in this patient (Epstein et al., 1987).

Both the EEGs and finding of LNNB suggest left temporal region of brain is more compromised. CT scan findings confirms it as a classical case of Dandy-Walker Syndrome (Epstein et al., 1987). In our case there was no cranial nerve abnormality and long tract signs, but seizures were the most important findings. Dandy-walker Syndrome is uncommon in adults, seizure are also very rare (Epstein et al., 1987); presence of post ictal psychosis is a new and unique finding which had not been reported yet. These constitute rarity of the presentation of Dandy-Walker Syndrome.

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S.C. PRADHAN, DPM. M.D. Senior Resident in Psychiatry. JNANAMAY DAS*, DPM, Resident in Psychiatry. VINOD KUMAR SINHA, DPM. M.D. Assistant Professor of Psychiatry. Central Institute of Psychiatry. Ranchi 834 006.

*Correspondence