A LARGE ASYMPTOMATIC DANDY WALKER CYST IN AN INFANTRY SOLDIER

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Introduction

Dandy Walker malformation is one of the causes of congenital hydrocephalus. It is cystic dilatation of fourth ventricle related to congenital absence of foramina of Magendie and Luschka, resulting in hypoplasia of cerebellum. In infants and young children, clinical features develop due to rising intracranial tension whereas in the older children the presentation depends upon the degree of neural agenesis manifesting mainly as a triad of dementia, gait ataxia and incontinence despite an apparently normal cerebro-spinal fluid (CSF) pressure. Long term outcome is dismal in most cases. Mortality remains high at 50-60% in children even with treatment [1]. However, 35% uncomplicated cases are expected to have a reasonable outcome [2]. We report an unusual presentation of a large Dandy Walker cyst in a serving infantry soldier who remained asymptomatic and was later detected only by MRI following a head injury.

Case Report

A 30 year old Sikh infantry soldier with twelve year service was admitted with history of having sustained head injury following involvement in assault during annual leave. On arrival, the patient was drowsy. No history of vomiting and seizures was available. The head was large, circumference 63 cm, with prominent occiput. Glasgow coma scale (GCS) was 9/15 (E2 V2 M5). Pupils were mid size and sluggishly reacting to light. MRI head showed a very large posterior fossa cyst of fourth ventricle, with symmetrical dilatation of lateral ventricles. Occipital lobes were compressed. Cerebellar vermis and corpus callosum were hypoplastic. All other lobes of brain were preserved in structure and myelination. The MRI findings were suggestive of Dandy Walker malformation (Figs 1,2,3). A small left parietal extradural haematoma was also present due to injury. The parietal haematoma was evacuated through a burr hole under local anaesthesia. Mannitol and glycerol were used for intracranial pressure reduction. Three weeks later, after recovery from effects of head injury, the patient was re-evaluated. Mild cerebellar ataxia along with mild constriction of visual field was now detected, but there was no papilloedema, or other cranial nerve sign. Intelligence quotient (IQ) assessed by performance tasks and verbal abilities was mildly re-

duced (64.5). Inquiry into his past performance in unit activities, training, discipline or other relevant medical history was inconclusive. The soldier had been a keen sportsman and achieved good grades in physical and weapon training events. He had also served in high altitude and operational areas without any noticeable disability. There had been no history of reporting sick with symptoms of rising intracranial pressure like severe headache, vomiting, altered consciousness. Due to this incidental finding of a large Dandy Walker malformation, a cysto-peritoneal shunt was done and the soldier was taken off combat duties.

Discussion

Dandy Walker malformation is a developmental anomaly in which presence of congenital septa or membrane causes obstruction to CSF outflow from the fourth ventricle thus creating a large cyst in the posterior fossa with symmetrical dilatation of ventricular system in brain. The large cystic expansion of the fourth ventricle results in agenesis of cerebellar vermis, corpus callosum and surrounding areas. Patients

Fig. 1 : MRI head mid sagittal image showing large posterior fossa cyst communicating with fourth ventricle. The cerebellar hemispheres and vermis are hypoplastic and displaced rostrally. Compression of occipital lobe is also seen.

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Fig. 2 : Axial image of same patient showing dilation of fourth ventricle and displacement of hypoplastic cerebellum

have large head, prominent occiput and other features of congenital hydrocephalus. Acute rise in intracranial pressure may cause papilloedema, altered sensorium and vomiting. Older children with normal pressure hydrocephalus have poor intelligence and memory, delinquent behaviour, cerebellar ataxia, abducent nerve palsy, visual field defects and pyramidal tract signs. Symptoms start manifesting in infancy till early adulthood, however, isolated cases have been reported to develop symptoms in late adult life [3]. Symptoms may often be related to previous trauma, meningitis or subarachnoid haemorrhage. Prognosis is linked to neurological impairment and associated developmental anomalies. 70% had subnormal IQ with poor memory in Swaya and McLaurin series [4]. Long term mortality in Fischer's series was 41%. Only 29% cases operated in infancy had near normal intelligence in long term multi disciplinary follow up to 14-22 years of age [5]. Neurological outcome in cases with isolated cerebellar hypoplasia was better than complicated cases [6].

The aim of reporting this case is that the detection of Dandy Walker malformation was an incidental finding at thirty years of age as the soldier had been asymptomatic probably due to near normal CSF pressures. The combat training and combat duties in infantry soldier require a high level of physical fitness, with good balancing, coordination of movements and sharp

Fig. 3 : Symmetrical dilatation of lateral ventricles with no midline shift

mental faculties. In addition, the soldiers are exposed to hostile terrains, altitudes, and vagaries of nature and demands of rigorous physical stress, which could precipitate symptoms. This soldier while serving in an infantry unit for twelve years, despite the presence of a large Dandy Walker cyst, was able to meet the demands of service without arousing suspicion even on routine half yearly medical examinations thus making it an unusual presentation.

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