A 15 years old boy presented with gradually developing enlargement of skull since childhood (Fig. 1). He has significant delay in physical and mental development. He has truncal ataxia. He has no history of vomiting and convulsions. He had no past history of meningitis or encephalitis. He has diminished power of both upper and lower limbs (4-/5) and has hypotonia. Jerks are preserved. Planter responses are bilaterally flexors. He has no sensory loss and cranial nerves are normal. Eye movements are normal. Cardiovascular, respiratory and gastroenterological system findings are normal. He has bilateral absence of 4th metatarsals (Fig. 2). Papilledema is absent. CT scan of brain shows that the 4th ventricle is replaced by an enlarged midline cyst. It is associated with aplasia of vermis, huge dilatation of other ventricles with thinning of the cortical mantle and absence of corpus callosum. All the black in the middle in Fig. 7 is water and the brain matter is the rim of white along the skull margin. The CT scan of brain is suggestive of Dandy-Walker Syndrome.

Dandy-Walker Syndrome is a genetically sporadic disorder that occurs 1 in every 25,000 live births, mostly in females. The primary defect was thought to be atresia of the foramina of Luschka and Magendie by Dandy and Blackfan (1914) and Taggart and Walker (1942). Benda (1954) introduced the designation Dandy-Walker Syndrome. He reported familial occurrences and described it a developmental anomaly not necessarily due to foraminal atresia since some cases had patent foramina. The definition of the syndrome proposed by Hart et al (1972) was three fold: 1. Partial or complete absence of cerebellar vermis, 2. posterior fossa cyst contagious with the 4th ventricle, and 3. Hydrocephalous. Hydrocephalous is a complication rather than part of the malformation complex. There is agenesis of midline cerebellum and large midline cyst, representing the greatly dilated 4th ventricle which occupies almost the entire posterior fossa. The tentorium; torcula, straight sinus, and vein of Galen are displaced superiorly. Occipital bone bulges posteriorly. It is frequently associated with disorders of other areas of CNS including absence of the corpus callosum and malformations of heart, face, limbs, fingures and toes (e.g., polydactyly, syndactyly). Hydrocephalus for obstruction almost always develops, but if treated promptly, the prognosis may be good.

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