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Neuropathological analysis of an asymptomatic adult case of Dandy-Walker Syndrome

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Case Report

The Dandy-Walker (DW) complex is a rare posterior fossa malformation, usually observed during the prenatal period or the early infancy. It consists of hypoplasia or agenesis of the cerebellar vermis, enlargement of the fourth ventricle with a posterior fossa cyst, and often hydrocephalus. The rare adult cases reported were characterized by late-onset symptoms, increased intracranial pressure and consistent presence of hydrocephalus. We report the first neuropathological analysis of an adult asymptomatic case of DW syndrome where the diagnosis was made incidentally, while searching for possible brain metastases of a breast tumor. An Ehlers-Danlos syndrome type IV characterized by repetitive intestinal perforations and a saccular aneurysm on the left posterior communicating artery was also present. Macroscopic examination revealed hypoplasia of both cerebellar hemispheres and posterior part of the vermis, dilatation of the IV ventricle without hydrocephalus and an arachnoid cyst in the posterior fossa. Histologically, there were two foci of cerebellar cortical dysplasia. In contrast, no ectopic neurons, neuronal loss or gliosis were observed in cerebral cortex and cerebellum. Mild neurofibrillary tangle formation was confined to the entorhinal cortex and CA1 field of the hippocampus. In agreement with the structural imaging data reported in adult DW cases, the neuropathological study of this case demonstrates the presence of milder brain abnormalities compared to DW infants. In this context, the preserved cortical cytoarchitecture and the rarity of additional neurodevelopmental changes may explain the absence of clinical expression