Dandy–Walker malformation associated with syringomyelia

Case illustration

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This 3-month-old boy was first assessed by a pediatrician for progressive macrocrania in October 1986. A computerized tomography scan revealed a Dandy–Walker malformation (DWM) with mild ventricular dilation. The patient was asymptomatic and therefore followed clinically and radiologically. At the age of 2 years, progressive hydrocephalus required cystoperitoneal shunt placement. Postoperative magnetic resonance (MR) imaging revealed progressive herniation of the lower pole of the cyst in the upper cervical canal (Fig. 1A) and the development of a syringomyelic cavity (Fig. 1B) that continued to increase in spite of three shunt revisions (Fig. 1C). The patient developed normally but the onset of gait disturbances prompted us to perform a suboccipital decompression, C-1 laminectomy, and duraplasty at the age of 9 years. An MR image obtained 3 months later (Fig. 1D) showed a significant decrease in the size of the syrinx.

The association between DWM and syringomyelia has rarely been described, either as isolated case reports or as an occasional finding in large autopsy or clinical series. Milhorat, et al., recently reported three cases of DWM with syringomyelia in a series of 105 autopsy specimens obtained in syringomyelia patients. The authors included these observations in the group that they called “communicating central canal syrinxes.” They stated that the occurrence of syringomyelia in DWM would be the expression of a participation of the central canal “in the hydrocephalus process like a fifth ventricle.”

This case seems to show that the cause of syringomyelia in DWM could be multifactorial. The crowding of the occipital foramen induced by the herniation of the lower pole of the cyst could be responsible for the progression of the syringomyelia, acting with a Chiari-like mechanism in spite of the presence of a working cerebrospinal fluid shunt. In such cases, suboccipital decompression and duraplasty are effective treatments.

References