Dandy Walker Syndrome and Associated Polycystic Renal Disease

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mation in patients with polycystic renal disease represents a fortuitous association or a component of the same process.

References

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Sir,

The Dandy-Walker syndrome is a congenital developmental disorder of the brain characterized by agenesis or hypoplasia of the cerebellar vermis and cystic dilatation of the fourth ventricle. The condition is usually associated with hydrocephalus depending on the permeability of the foramina of Magendie and Luschka [1,2]. The clinical manifestations range from none to severe disturbances of development including signs of cerebral compression and mental retardation. Sporadically, it has also been described as a component of malformation syndromes in infants [3, 4]. We report a case of Dandy-Walker syndrome in an adult patient with polycystic renal disease.

A 30-year-old woman was admitted to the emergency room because of hypovolemic shock caused by severe metrorrhagia. She suffered from chronic renal failure due to polycystic renal disease and was maintained on hemodialysis therapy since the age of 20; on the seventh hospital day she presented a generalized tonic-clonic seizure. The patient recovered with standard pharmacotherapy and the subsequent neurological examination did not disclose abnormalities. The EEG showed desynchronization and slow waves in the right temporal areas; a CT scan showed a cystic dilatation of the posterior fossa in wide-open communication with the fourth ventricle, without hydrocephalus. The NMR imaging confirmed (fig. 1) previous CT findings and the evidence of a hypoplastic vermis. Polycystic renal disease was confirmed by abdominal ultrasound and liver cysts were not detected. Ct and NMR sections may show, in the future, if Dandy-Walker malfor-

Fig. 1. NMR showing a cystic dilatation of the posterior fossa in communication with the IVth ventricle and hypoplastic vermis.

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