Surgical management and clinical results in a series of interhemispheric arachnoid cysts

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Background and Objective: Arachnoid cysts (AC) located within the interhemispheric fissure are rare, accounting for <5% of all cases, and frequently associated with agenesis or hypogenesis of the corpus callosum.

They consist of a heterogeneous group of disorders that Barkovich classified in two major types, each with subtypes, based on morphology, ventricular communication, and association with other cerebral malformations. Mori further classified these cysts as either intra- or extraxial depending on their location associated ventricular communication and corpus callosal agenesis.

Optimal surgical management of these cysts and its impact on neurological development is controversial and matter of debate. Surgical options consist of cyst fenestration, either cystoventriculostomy or cystocisternostomy or both, or placement of a shunt, but surgical suggestions are based on case reports or small series.

We present our surgical experience and its clinical results in children with large interhemispheric AC who underwent cystoperitoneal shunting (CP) and evaluated the long-term outcome, on clinical data, psychomotor development and neuroimaging.

Series: Four patients (three males, one female) were treated at our institution from 2005 to 2015 for interhemispheric arachnoid cysts. All patients were symptomatic with macrocrania, intracranial hypertension, and epileptic seizure in one case. Brain MRI showed interhemispheric (multiloculated in two patients) cyst of CSF density, with no-enhancing cystic wall, in association with complete (one case) or partial agenesis of the corpus callosum.

All the cysts were extraxial noncommunicating interhemispheric following the Mori Classification, Type 2a in two children, 2b in one and 2c in one according to the Barkovich Classification.

Despite the diagnosis has been perinatal in all cases, the age at surgery was 2, 7, 44 and 76 months, due to delayed neurosurgical referral. Consequently all the cysts were large, occupying more than one third of the supratentorial space at time of surgery.

Results: All patients underwent cystoperitoneal shunting, with multi-perforated catheter and endoscopic assistance; in one case a fenestration was associated. Programmable shunts were applied, progressively changing the opening pressure based on neuroimaging cyst shrinkage. The surgical choice was based on the huge dimension of the cysts.

The patients were followed up for an average of 36 months (range 11-73 months). Postoperative Neuroimaging showed a reduction of cyst in all cases. The major parenchymal re-expansion was obtained in the 2 months girl.

The clinical outcome was good with complete resolution of symptoms and signs. The developmental outcome scale, which included cognitive and psychomotor development, was normal for three patient and slightly delayed in one.
Conclusions: The present series of large interhemispheric cysts, lately treated, had good results in terms of cerebral parenchymal re-expansion with programmable cystoperitoneal shunts. This reflected in a favorable clinical outcome, achieved both in the short and the long term.

Even if the optimal surgical treatment for this type of cysts is probably neuro-endoscopic early fenestration, the endoscopic assisted shunting is a reasonable alternative in case of long standing huge cysts because it offers the chance of a progressive decrease of parenchymal compression that favors its re-expansion.