Hydrocephalus after Diffuse intrinsic pontine glioma

Brainstem glioma accounts for 6-9% of brain tumors in children. Tumor progression may lead to CSF pathway obstruction and development of hydrocephalus.

Diffuse Intrinsic Pontine Gliomas (DIPG) related hydrocephalus occurs as the result of tumor growth and aqueductal stenosis. There is no consensus about the best surgical option, thus a review has been performed to clarify the rate of success, complications and possible issues of Endoscopic Third Ventriculostomy (ETV) in comparison to the other available techniques.

A systematic review followed the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) statement and it was registered with the PROSPERO International Prospective Register of Systematic Reviews(CRD42018089001). MEDLINE, Web of Science and EMBASE were searched for published series in which ETV was performed to treat hydrocephalus in DIPG patients.

Six studies were included. Two further cases coming from the experience of Guida et al., were added, for a total amount of 55 patients treated through either ETV, VPS or Ventriculocisternal shunt according to Arne Torkildsen. 86% of patients who underwent ETV experienced clinical improvement after surgery (p-value 0.03). Torkildsen shunt placement was associated to 50% failure rate. Two patients implanted with VPS developed symptoms of shunt malfunction and increased ventricular sizes (10%). Fisher's exact test was applied to compare efficacy of VPS and ETV with no statistical difference between the two group (p-value 0,17). Patients who underwent ETV did not experienced major complications and no procedural abortion was observed.

ETV is an effective and safe treatment option, associated to a low complications rate and a high success rate. Evidences from this review suggest to consider ETV as the first line treatment of hydrocephalus in DIPG patients [1)].

Roujeau et al., retrospectively reviewed charts of patients consecutively treated in Necker-Enfants Malades Hospital with diffuse intrinsic pontine glioma in order to assess incidence of hydrocephalus, its management, and its impact on overall survival. All patients had brain stem glioma not amenable to surgery. Cases with exophytic brain stem glioma were excluded.

Fifty-one children were treated from January 2005 to December 2010 for brain stem glioma in the Pediatric Neurosurgery Department of Necker Enfants Malades, Paris, France. Hydrocephalus occurred in 11 of them (22%). They were six boys and five girls; the average and median time from tumor diagnosis to onset of hydrocephalus were 5.3 and 3.2 months, respectively, while average and median time from onset of hydrocephalus to death were 5.3 and 2.8 months, respectively. Hydrocephalus was treated in nine patients by a ventriculoperitoneal (VP) shunt and in two patients by an endoscopic third ventriculostomy. Because of early failure, a VP shunt was implanted in one child.

The overall 1-year survival rate was 33%. Survival rate of patients with such obstructive hydrocephalus was not significantly different from patients harboring brain stem glioma who did not

develop hydrocephalus. Furthermore, hydrocephalus was not related to terminal tumor progression. Considering both risks and benefit of treatment, VP shunt could be proposed, on the base of our experience, as the first option in spite of the apparently obstructive nature of the hydrocephalus associated to a brain stem tumor [2].

References

1)

Guida L, Roux FE, Massimino M, Marras CE, Sganzerla E, Giussani C. Safety and efficacy of Endoscopic Third Ventriculostomy in Diffuse Intrinsic Pontine Glioma related hydrocephalus: a Systematic Review. World Neurosurg. 2018 Dec 29. pii: S1878-8750(18)32919-X. doi: 10.1016/j.wneu.2018.12.096. [Epub ahead of print] Review. PubMed PMID: 30599251.

2)

Roujeau T, Di Rocco F, Dufour C, Bourdeaut F, Puget S, Rose CS, Zerah M. Shall we treat hydrocephalus associated to brain stem glioma in children? Childs Nerv Syst. 2011 Oct;27(10):1735-9. doi: 10.1007/s00381-011-1538-2. Epub 2011 Sep 17. PubMed PMID: 21928037.