Spinal ependymomas in NF2: a surgical disease?


The management of spinal cord ependymomas in Neurofibromatosis Type 2 (NF2) has traditionally been conservative, in contrast to the management of sporadic cases; the assumption being that, in the context of NF2, they did not cause morbidity. With modern management and improved outcome of other NF2 tumours, this assumption, and therefore the lack of role for surgery, has been questioned. To compare the outcome of conservative treatment of spinal ependymomas in NF2 with surgical intervention in selected patients.

Retrospective review at two NF2 centers, Manchester, UK and Paris/Lille, France. In Manchester patients were managed conservatively. In France surgery was a treatment option. Inclusion in the study was based on tumor length of greater than 1.5 cm. The primary parameter assessed was acquired neurological deficit measured by the Modified McCormick Outcome Score. 24 patients from Manchester and 46 patients from France were analyzed. From Manchester, 27% of these patients deteriorated during the course of follow-up. This effectively represents the natural history of ependymomas in NF2. Of the surgical cases, 23% deteriorated postoperatively, but only 2/18 (11%) of those operated on in the NF2 specialist centers. Comparison of the two specialist centers Manchester/France showed a significantly improved outcome (P = 0.012, χ² test) in the actively surgical center. Spinal ependymomas produce morbidity. Surgery can prevent or improve this in selected cases but can itself can produce morbidity. Surgery should be considered in growing/symptomatic ependymomas, particularly in the absence of overwhelming tumor load where bevacizumab is the preferred option.

KEYWORDS: Ependymoma; McCormick grading system; NF2; Neurofibromatosis type 2; Spinal cord

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