Management of Ganglioneuroma and Ganglioneuroblastoma Intermixed A UNited Kingdom Childrens cancer and Leukaemia Group (UKCCLG) Nationwide Study REPORT

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Abstract

Background Ganglioneuroblastoma intermixed (GNBi) and ganglioneuroma (GN) are benign subtypes of neuroblastic tumors. Primary observation has become accepted management for some patients with surgical operative strategies evolving to be less aggressive. Objectives Our study examines evolving management in a UK cohort investigating natural history, biology and clinical features of GN and ganglioneuroblastoma-intermixed (GNBi) in those having observation or surgery. Methods Retrospective review of histologically confirmed GN and GNBi managed over a 30 year period. Clinical, pathological features, tumor dimensions, management and outcomes are all recorded. Results A total of 259 patients were identified (GN= 163, GNBi = 93, median age = 62 months). 201(78%) had upfront surgery and 58 (22%) were actively observed. Of the 58 observed - 21 (36%) later required surgery due to progressive tumour growth (52%). Gross total resection was achieved in 79% of patients with a 19% complication rate. Presence of image defined risk factors and large tumour size correlated with incomplete resection ($p < 0.05$ in both). Forty-five index cases (39%) had change in pathology between biopsy and surgery with 14 patients (12%) altered from 'favourable' to 'unfavourable'. Conclusion Our findings show surveillance alone may be considered a safe approach. However, a significant number of index patients may eventually require operative surgery with development of symptoms.

Extent of surgical resection did not impact overall survival (OS); however it improved symptom(s) resolution.

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