Nelson's syndrome treatment with Gamma knife radiosurgery

The best treatment for Nelson's syndrome has not been well defined. Gamma knife radiosurgery (GKRS) is very effective to stop the growth of the pituitary neuroendocrine tumor, which is the main goal of the treatment of patients with NS.

Losa et al. reported the largest series of patients with NS treated by GKRS at a single center.

The study was an observational, retrospective analysis of 28 consecutive patients with NS treated by GKRS in the department between 1995 and 2019. All patients had a growing Pituitary corticotroph adenoma. The main outcome of the study was to assess by the Kaplan-Meier method the risk of tumor progression after GKRS.

The median follow-up after GKRS treatment was 98 months (IQR 61-155 months, range 7-250 months). Two patients (7.1%) had a recurrence of the disease during follow-up. The 10-year progression-free survival was 91.7% (95% CI 80.5-100%). No patient had deterioration of visual function or oculomotor function after GKRS. New onset of hypogonadism and hypothyroidism occurred in 18.8% and 14.3% of the patients at risk.

The study confirms that GKRS may stop the tumor growth in the majority of patients with NS, even though very aggressive adenomas may ultimately escape this treatment. Safety of GKRS was good in our experience, but due attention must be paid to planning the distribution of radiation to critical structures, especially in patients previously treated by radiation [1)].

In a study Cordeiro et al., sought to better define the therapeutic role of stereotactic radiosurgery (SRS) in Nelson's syndrome.

Study patients with Nelson's syndrome were treated with single-fraction GKRS (median margin dose of 25 Gy) at 6 different centers as part of an International Radiosurgery Research Foundation (IRRF) investigation. Data including neurological function, endocrine response, and radiological tumor response were collected and sent to the study-coordinating center for review. Fifty-one patients with median endocrine and radiological follow-ups of 91 and 80.5 months from GKRS, respectively, were analyzed for endocrine remission, tumor control, and neurological outcome. Statistical methods were used to identify prognostic factors for these endpoints.

At last follow-up, radiological tumor control was achieved in 92.15% of patients. Endocrine remission off medical management and reduction in pre-SRS ACTH level were achieved in 29.4% and 62.7% of patients, respectively. Improved remission rates were associated with a shorter time interval between resection and GKRS ($p = 0.039$). Hypopituitarism was seen in 21.6% and new visual deficits were demonstrated in 15.7% of patients.

GKRS affords a high rate of pituitary neuroendocrine tumor control and improvement in ACTH level for the majority of Nelson's syndrome patients. Hypopituitarism is the most common adverse effect from GKRS in Nelson's syndrome patients and warrants longitudinal follow-up for detection and endocrine replacement [2)].

Twenty-seven patients with Nelson's syndrome treated with Gamma Knife radiosurgery after bilateral adrenalectomy were included in this study. After radiosurgery, patients were followed with serial adrenocorticotropic hormone (ACTH) levels and MRI sequences to assess for endocrine remission and tumor control. Cox proportional hazards regression analysis was used to evaluate the relationship between the time to remission and potential prognostic factors.

Results: In 21 patients with elevated ACTH prior to SRS and endocrine follow-up data, 14 (67%) had decreased or stable ACTH levels, and 7 achieved a normal ACTH level at a median of 115 mo (range 7-272) post-SRS. Tumor volume was stable or reduced after SRS in 92.5% of patients (25/27) with radiological follow-up. Time to remission was not significantly associated with the ACTH prior to SRS (P = .252) or with the margin dose (P = .3). However, a shorter duration between the patient's immediate prior transsphenoidal resection and SRS was significantly associated with a shorter time to remission (P = .045).

Conclusion: This retrospective analysis suggests that SRS is an effective means of achieving endocrine remission and tumor control in patients with Nelson's syndrome [3].

1)
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2)
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