

Surgical Management of MRI Negative Cushing Disease

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Introduction

Management of patients with MRI-negative Cushing's disease (CD) can be challenging; factors predicting tumor localization remain imperfect. Reported remission rates in the literature vary widely (~60 to 86%), further complicating decision-making. We examined our experience in 52 patients, over 7.5 years, operated upon by a single surgeon.

Learning Objectives

- 1) IPSS data must be interpreted with an understanding of its limitations
- 2) Prolactin-adjusted ACTH levels can improve the predictive value of IPSS in laterality prediction

Methods

Prospectively-collected data of 52 patients with MRI-negative CD were reviewed retrospectively. All patients who underwent transphenoidal surgery by a single surgeon from March 2006 to September 2013 were analyzed. Of these, nine (17.3%) had prior surgery elsewhere, without remission or tumor identification.

Results

A tumor was identified at surgery in 47/52 (90%) patients; an ACTH-staining pituitary adenoma was confirmed in 38/47 (81%) of these tumors. Initial remission was achieved in 44/52 (84.6%) cases, with recurrence in 4/44 (9.0%) patients (mean time to recurrence 40.5 months, range 6-87). Excluding prior surgery patients, remission was achieved in 38/43(88%) patients with a mean follow-up of 34 months (range 1-118); there were three recurrences (3/38 or 7.8%). IPSS was performed in all patients; in 28/52 (53.8%) patients tumors lateralized; in 19 (67.8%), IPSS lateralization correlated with the location of the adenoma. Prolactin-adjusted ACTH ratios were more predictive of laterality than the unadjusted ACTH ratio (p=.044). Excluding four cases of hemi- or complete hypophysectomy, new postoperative hormone deficits were noted in 4/48 (8.3%) patients. Mean follow-up time was 37.5 months (range 1-118).

Conclusions

Transphenoidal exploration and resection of radiographically negative CD can result in a high rate of tumor identification and durable remission with a low rate of new postoperative deficits. 88% of Cushing's patients undergoing their first surgery achieved remission at a mean follow-up of 34 months. While IPSS may assist in guiding resection in MRI-negative cases, it remains an imperfect indicator of tumor laterality. The use of prolactin-adjusted levels may enhance the predictive value of IPSS.