Diagnosis and Management of Cushing Disease

WILLIAM T. COULDWELL, M.D., PH.D.

Department of Neurosurgery, University of Utah, Salt Lake City, Utah

The management of a case involving Cushing disease, defined as excess secretion of adrenocorticotropic hormone (ACTH) from a pituitary adenoma, remains one of the most vexing problems facing the pituitary surgeon. The systemic consequences of this endocrinopathy are profound morbidity and premature death in the unfortunate patients in whom remission fails to occur. For this reason, the challenge to endocrinologists and neurosurgeons charged with treating these patients is normalization of pathological ACTH and, thus, serum cortisol levels. Unfortunately, transsphenoidal surgery fails to produce remission of symptoms in a significant percentage of patients, and repeated surgery or the institution of adjuvant therapy must be considered in such cases.

In this issue of Neurosurgical Focus, a series of papers has been selected to provide a comprehensive overview of the diagnosis and management of this complicated disease. Papers involving the contemporary diagnostic workup and pitfalls, as well as success rates and outcomes following transsphenoidal surgery, are presented. Authors present various treatment options for patients in whom initial surgery has failed to resolve symptoms. A fascinating review paper on the phenomenon of brain atrophy seen in Cushing disease and its possible pathogenesis is included. Finally, two papers—one a historical overview and the other a contemporary review of the management of Nelson syndrome, a dreaded complication of an aggressive ACTH-secreting tumor of the pituitary developing after bilateral adrenalectomy—are presented.

I would like to acknowledge the efforts of Margie Shreve and Dr. Martin Weiss. Without their expert and timely help, compiling this issue would not have been possible.

(DOI: 10.3171/FOC-07/09/EIntro)