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Introduction

Ectopic adrenocorticotrophic hormone (ACTH-ectopic) syndrome (EAS) is a rare cause of ACTH-dependent endogenous hypercortisolism. The objective of this study was to analyze the clinical, biochemical, and radiological features, management, and treatment outcome of patients with EAS.

Materials and methods

It was a retrospective case-record study of 52 patients with EAS. Clinical, biochemical, and radiological features and response to therapy and survival were measured.

Results:

The median follow-up was 7 yr (range, 1–13 yr). None of the dynamic tests achieved 100% accuracy. Imaging correctly identified the lesion at first investigation in 83% of cases. Bronchial carcinoid tumors were the most common cause of EAS (n = 34; 65.4%), followed by other neuroendocrine tumors (n = 13, 25%). In 9.6% (5) of patients, the source of EAS was never found. Octreotide scintigraphy and whole-body venous sampling were of limited value. Surgical attempt at curative resection was successful in 83% (43 of 52) of all patients; 9 (19.1%) responded generally well to bilateral adrenalectomy by vital necessity. Tumor histology and the presence of distant metastases were the main predictors of overall survival (P < 0.05).

Conclusion

No single test provides to find the source of EAS correctly. Despite a variety of tests and imaging studies for the correct diagnosis of the EAS, up to 10% of cases present an occult EAS syndrome. These cases require a prolonged follow-up, review, and repetition of diagnostic tests and scans, but, if it is necessary, do bilateral adrenalectomy.

References

Ma Y, Altei C, Dobson RW, Konduri K. Ectopic adrenocorticotropic hormone syndrome: a diagnostic challenge and review of the literature.//Proceedings (Baylor University, Medical Center) – 2010 – Vol. 23 – P. 426–428