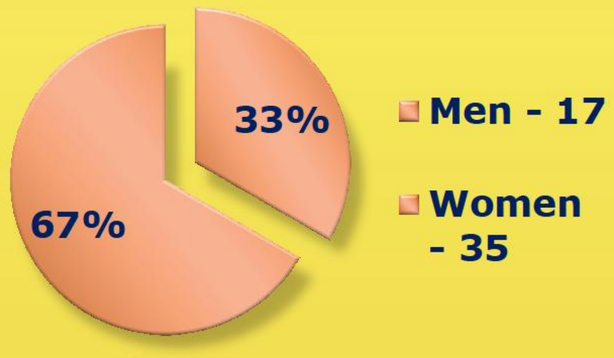
EP-650: Ectopic ACTH syndrome: Clinical features, diagnosis, treatment and observation.

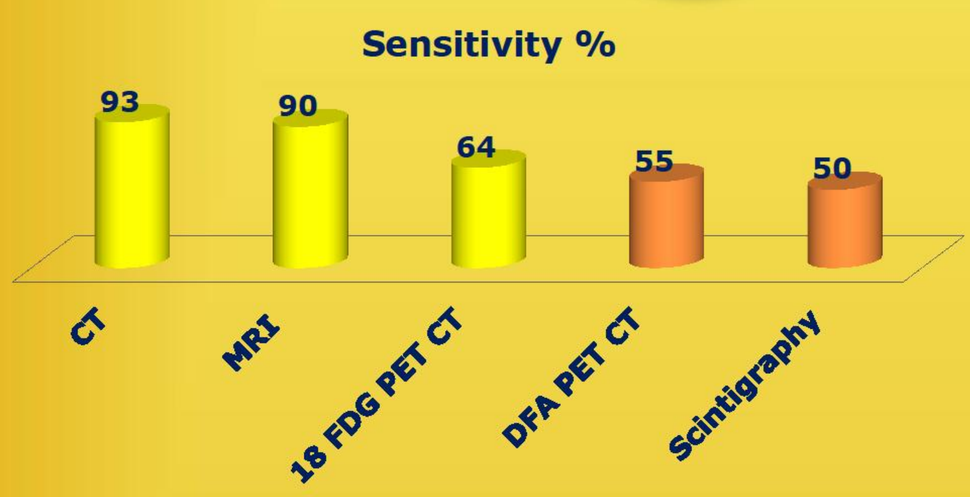
Krylov V. , Dobreva E., Kuznetsov N., Marova E., Ippolitov L., Latkina N.
Department of Endocrine Surgery
Endocrinology Research Center Moscow, Russia

52 patients with ectopic ACTH syndrome

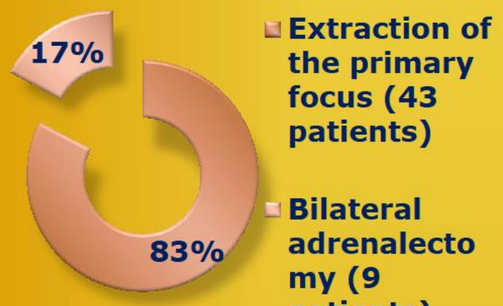
The average age of 41 ± 16,5 years

The average duration of the disease 2,7 ± 1,6 years

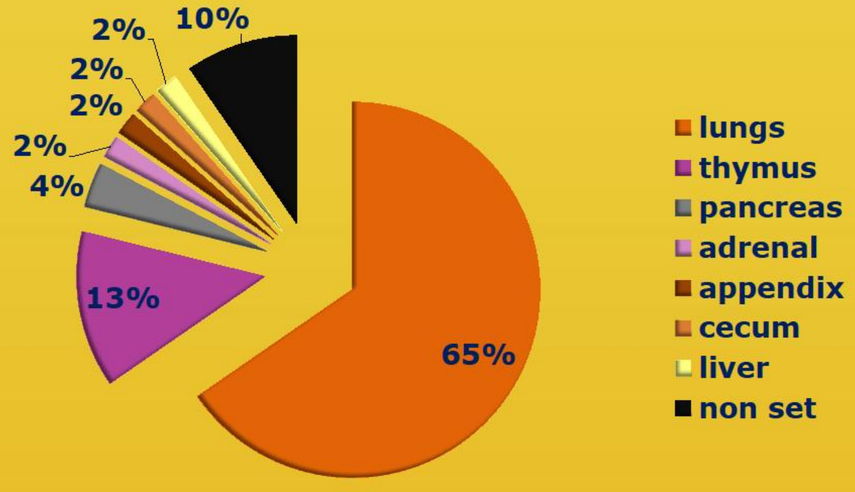




Surgical treatment

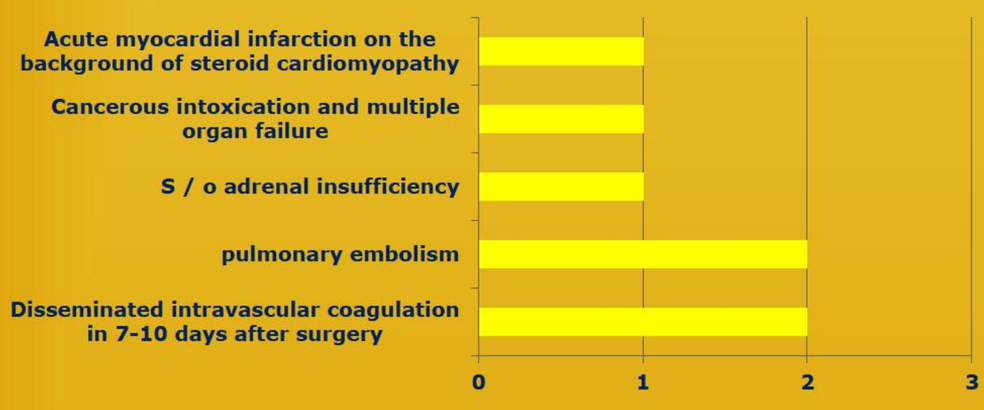


Localization of the primary focus Σ = 52 cases

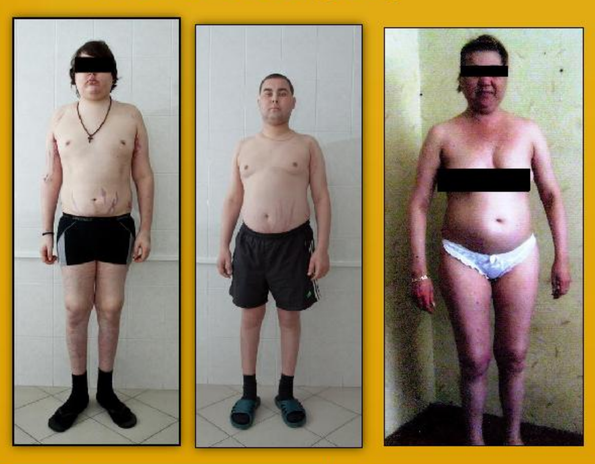


Deaths Σ = 7

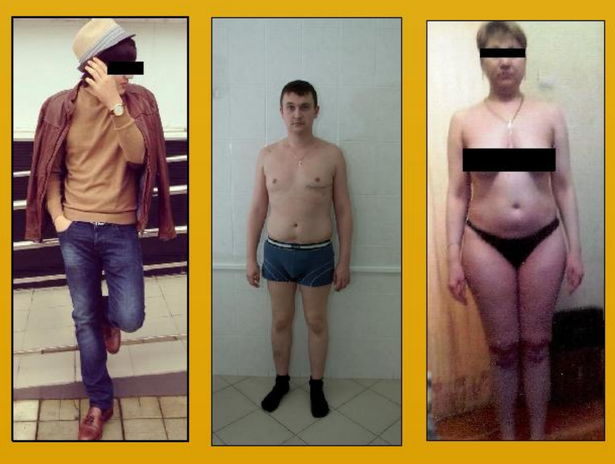
Cause of deaths Σ = 7



Before surgery



6 months. after surgery



Introduction

Ectopic adrenocorticotropic 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

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