ADDISONIAN CRISIS AS A MANIFESTATION OF A PARTIALLY EMPTY SELLA IN A 68-YEAR-OLD WOMAN

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OBJECTIVES
Empty sella syndrome (ESS) is a condition often discovered incidentally, where the sella turcica, the structure containing the pituitary gland, appears to be empty.

Patients either remain asymptomatic or, rarely, manifest signs of declined pituitary function. Patients experiencing hypopituitarism are offered hormonal replacement.

Autopsy studies estimated a 5% EES prevalence among healthy individuals.

METHODS
Case report: A 68-year-old post-menopausal woman, overweight (BMI=29), mother of 3 children, presented with a 3-day history of fever (T=38, 5°C), excessive vomiting and nausea and a history of 15 days of anorexia, weakness and fatigue.

Five years ago, during the course of a hospitalization for the investigation of leukenopia, she was diagnosed with myelodysplastic syndrome and received treatment with pegfilgrastin and epoetin beta.

RESULTS
Physical examination yielded no pathological findings, whereas laboratory results revealed leukenopia (WBC=2.900), hypokalemia (K+ 3.2 mmol/L), low TSH levels=0.18 μIU/ml, low FT4=6.01 pmol/L, n.v. 12.0-22.0 pmol/L), decreased gonadotropin levels (FSH=1.3 μIU/ml, n.v. in menopause >30.0 μIU/ml and LH=0.2 μIU/ml, n.v. in menopause >10.0 μIU/ml), hypocortisolemia (cortisol=51.0 nmol/L, n.v. 171.0-536.0 nmol/L) and low normal ACTH=17.3 pg/ml (n.v. 10.0-60.0 pg/ml).

The patient underwent adrenal function investigation with the short Synacthen test, where she responded positively (cortisol 0 min=110.0 nmol/L, 30 min=283.0 nmol/L, 60 min=410.0 nmol/L).

MRI of the pituitary gland demonstrated a partially empty sella turcica, with herniation of the suprasellar cistern.

The patient was diagnosed with panhypopituitarism and was initially treated with i.v. administration of methylprednisolone.

After discharge, outpatient treatment included levothyroxine and prednisolone.

CONCLUSIONS
Empty sella may be primary or secondary to surgery, irradiation or infarction of the pituitary gland. Idiopathic ESS usually generates from congenital defects of the sellar diaphragm where arachnoid membrane herniates through the deficient diaphragm, compressing the pituitary gland. In up to 50% of the cases, primary ESS is associated with benign intracranial hypertension.

MRI usually demonstrates the compression of the pituitary tissue against the floor of the sella and the subsequent deviation of the pituitary stalk. Hypopituitarism manifests when >90% of the pituitary tissue is compressed or atrophied.