CRANIOPHARYNGIOMAS – 35 YEARS OF EXPERIENCE IN A CENTRAL HOSPITAL'S ENDOCRINOLOGY DEPARTMENT

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INTRODUCTION

- Craniopharyngiomas: rare epithelial tumors of the sellar and parasellar region, with high survival rates (~90%) but with frequent tumor recurrence or persistence and significant morbidity.
- Annual incidence: 0.5-2.0 cases/million/year.
- Bimodal age distribution: peak incidence rates at 5–14 years and at 50–74 years.
- Clinical manifestations related to mass effect and hypothalamic/ pituitary deficiencies.

METHODS

- Information collection from clinical records
- Review of the epidemiology, diagnosis, treatment and follow-up of patients with diagnosis of craniopharyngioma followed in an Endocrinology Department between 1980 and 2015
- Statistical analysis using SPSS v. 22.0

RESULTS

- Sample size: **40 patients**
- 50% male, 50% female
- Median age by diagnosis: **36 years (minimum 6, maximum 70)**
- **Clinical manifestations at diagnosis:**

  visual alterations 77,5% (n=31)

  headache 72,5% (n=29)

- 77,5% with **sellar + suprasellar involvement**
- **Cystic component** detected in 89,7% and **calcifications** in 47,5%
- Initial surgical approach: **transcranial** in 74,4%, transsphenoidal in 25,6%
- Neuropathology results (n=29):

  79,3% (n=23) **adamantinomatous**

  20,7% (n=6) papillary

- Average number of surgical interventions per patient: **2 (minimum 1, maximum 7)**
- Eight patients (20.0%) treated with radiotherapy
- 47,5% of patients **with residual tumor** in the last neuroimaging evaluation

- **Panhypopituitarism** detected in 6/30 patients preoperatively and in 23/39 in follow-up
- All isolated anterior pituitary deficits also more frequent in follow-up

	At diagnosis	Last follow-up
Panhypopituitarism	20,0% (n=6/30)	**59,0%** (n=23/39)
GH deficit	67,7% (n=21/31)	**74,4%** (n=29/39)
Secondary hypogonadism	81,3% (n=26/32)	**85,0%** (n=34/40)
Secondary hypothyroidism	40,0% (n=12/30)	**82,5%** (n=33/40)
Secondary adrenal insufficiency	38,7% (n=12/31)	**75,0%** (n=30/40)

- Weight gain seen in **83,9%** of patients **(average ↑ 20,5 Kg)**
- Prevalence of diabetes mellitus, arterial hypertension and dyslipidemia also increased

	At diagnosis	Last follow-up
Overweight/ obesity	56,2% (n=18/33)	**87,1%** (n=27/31)
Dyslipidemia	36,8% (n=14/38)	**62,5%** (n=25/40)
Arterial hypertension	13,2% (n=5/38)	**20,0%** (n=8/40)
Diabetes mellitus	5,3% (n=2/40)	**12,5%** (n=5/40)
Central diabetes insipidus	16,2% (n=6/37)	**57,5%** (n=23/40)

- Average follow-up time: 15,5 (± 10) years
- Six patients died (4 diagnosed between 6 and 14 years of age)
- Neurological sequelae: 32,4% (n=11/34)
- Visual deficits: improvement in 51,4% (n=19/37), unchanged in 45,9% (n=17/37)

CONCLUSION

- Despite the evolution in neurosurgical technique and the possibility of radiotherapy, craniopharyngiomas are still associated with high rates of tumor persistence and substantial morbidity.
- Hypothalamic alterations, pathological or treatment-induced, have a major impact on prognosis mainly due to hypothalamic obesity. Treatment decisions for primary and recurrent disease need to consider long-term tumor control as well as treatment-related morbidity.
- Craniopharyngioma often must be managed as a chronic disease by experienced multidisciplinary teams.
- Optimization of the treatment of any endocrine and metabolic sequelae is essential.

References: Erfurth EM *et al*, *Mortality and morbidity in adult craniopharyngioma*, Pituitary 2013;16(1):46-55. Müller HL, *Craniopharyngioma.* Endocr Rev 2014;35(3):513-43. Nielsen EH *et al*, *Incidence of craniopharyngioma in Denmark (n = 189) and estimated world incidence of craniopharyngioma in children and adults,* J Neurooncol 2011;104(3):755-63.