

CRANIOPHARYNGIOMA – A DIAGNOSIS NOT TO BE MISSED



Joana Menezes 1,2, Elisabete Rodrigues 1,2, Sérgio Salvador 3, António Cerejo 3, Ricardo Reis 4, Sérgio Silva 4, Luís Augusto 5, Marcos Guimarães 6, Rui Vaz 2,3, Davide Carvalho 1,2

1 – Endocrinology, Diabetes and Metabolism Department, 2 – Faculty of Medicine University of Porto, 3 – Neurosurgery Department, 4 – Ophtalmology Department, 5 – Neurorradiology Department, 6 – Pathology Department

CENTRO HOSPITALAR SÃO JOÃO

INTRODUCTION

- ✓ Craniopharyngiomas are intracranial tumors that develop from epithelial Rathke's pouch rests. They have a bimodal age distribution, with peak incidence at childhood and in the adult/elderly, although in our series we have more patients in a middle peak.
- ✓ Usually they are benign but responsible for significant morbidity, particularly when located near critical structures such as optic chiasm, pituitary gland and hypothalamus, and thus might cause visual, neurological and endocrine deficits.

Jan/11 Oct/11

24-year-old male, healthy

Progressive visual impairment of the right eye until amaurosis

Without other neuroophthalmological changes neither hormonal dysfunction symptoms. HORMONAL EVALUATION: subclinical hypothyroidism (started on levothyroxine therapy 50μg/day).

PITUITARY MRI: sellar and suprasellar cystic lesion with carotid involvement. This lesion was not completely excised, because of its adherence to internal carotid.

HISTOLOGY: craniopharyngioma adamantinomatous type.

1st Endocrinology Consultation

HYPOTHYROIDISM
CENTRAL DIABETES
INSIPIDUS

Levothyroxine and desmopressin therapy (0.2mg/day).

2nd Endocrinology Consultation PANHYPOPITUITARISM

Nov/11

Requiring adjustment of levothyroxine besides testosterone and prednisone supplementation.

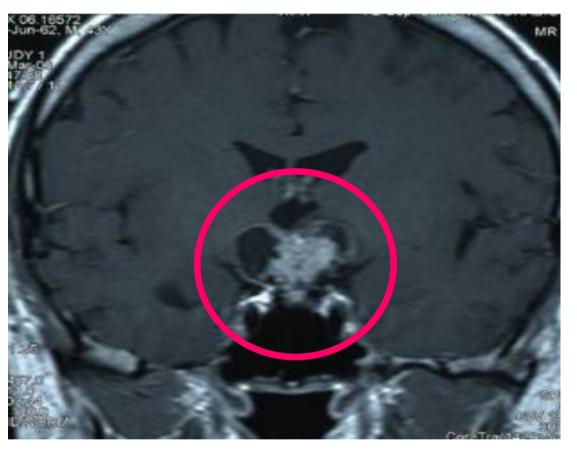
PITUITARY FOLLOW-UP MRI:

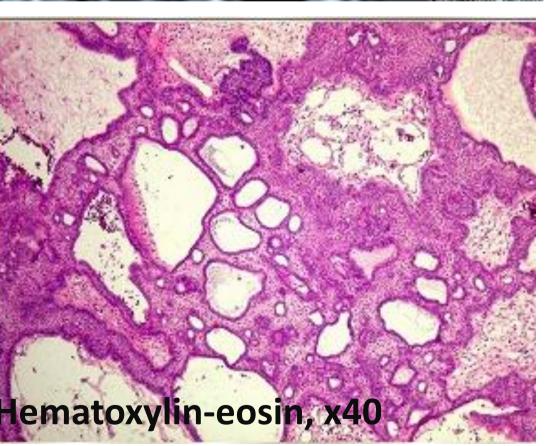
Sequelae of surgery, tumour-adjacent residues and optic chiasm with scoop bottom and with discreet focus of the contrast on the inferior right ocation.

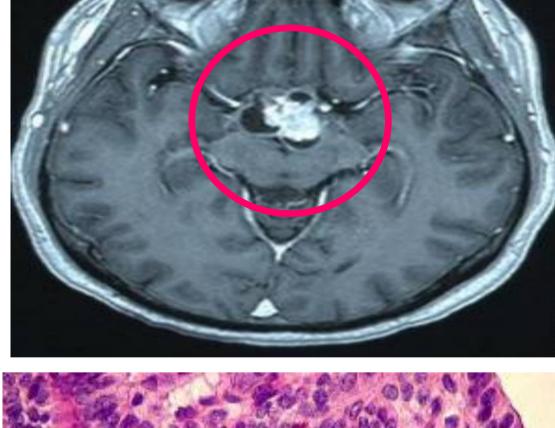
CLINICALLY:

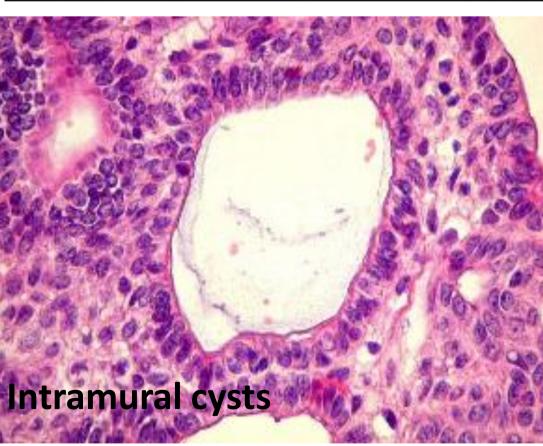
Visual impairment of the right eye (<1/10), relative afferent pupillary defect, and pallor of optic disc in ophthalmoscopy, without other neurological or endocrine symptoms.

	Value	Normal Range
TSH	0.20 mUI/mL	0.35-4.94
FT4	0.73 ng/dL	0.70-1.48
FT3	1.91 ng/dL	1.71-3.71









CRANIOPHARYNGIOMA ADAMANTINOMATOUS TYPE Complex epithelial lesion with several distinctive morphologic features: peripheral palisading of the epithelium; frequently, the inner epithelium beneath the superficial palisade undergoes hydropic vacuolization as is referred to as the stellate reticulum; Intramural cysts; scattered nodules of keratin

Value Normal Range FT4 0.44 ng/dL 0.70-1.48 FT3 2.82 ng/dL 1.71-3.71 TSH 3.05 μUI/mL 0.35-4.94 Total Testost <0.03ng/mL 2.8-8.0 LH 0.48 mUI/mL 1.7-8.6 FSH 0.97 mUI/mL 1.5-12.4 Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300 Urinary Osmol 407 mOsmol/kg 50-1200			
FT3 2.82 ng/dL 1.71-3.71 TSH 3.05 μUI/mL 0.35-4.94 Total Testost <0.03ng/mL 2.8-8.0 LH 0.48 mUI/mL 1.7-8.6 FSH 0.97 mUI/mL 1.5-12.4 Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300		Value	Normal Range
TSH 3.05 μUI/mL 0.35-4.94 Total Testost <0.03ng/mL 2.8-8.0 LH 0.48 mUI/mL 1.7-8.6 FSH 0.97 mUI/mL 1.5-12.4 Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300	FT4	0.44 ng/dL	0.70-1.48
Total Testost <0.03ng/mL 2.8-8.0 LH 0.48 mUI/mL 1.7-8.6 FSH 0.97 mUI/mL 1.5-12.4 Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300	FT3	2.82 ng/dL	1.71-3.71
LH 0.48 mUI/mL 1.7-8.6 FSH 0.97 mUI/mL 1.5-12.4 Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300	TSH	3.05 μUI/mL	0.35-4.94
FSH 0.97 mUI/mL 1.5-12.4 Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300	Total Testost	<0.03ng/mL	2.8-8.0
Prolactin 20 ng/mL 4.0-15.0 Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300	LH	0.48 mUI/mL	1.7-8.6
Cortisol 0.43μg/dL 6.2-19.4 ACTH 11.6 ng/L <63.3 Serum Osmol 285 mOsmol/kg 282-300	FSH	0.97 mUI/mL	1.5-12.4
ACTH 11.6 ng/L <63.3	Prolactin	20 ng/mL	4.0-15.0
Serum Osmol 285 mOsmol/kg 282-300	Cortisol	$0.43 \mu g/dL$	6.2-19.4
	ACTH	11.6 ng/L	<63.3
Urinary 0smol 407 mOsmol/kg 50-1200	Serum Osmol	285 mOsmol/kg	282-300
	Urinary 0smol	407 mOsmol/kg	50-1200

CONCLUSION

✓ Craniopharyngiomas are rare tumours, often with suprasellar extension. Early diagnosis and treatment require a high diagnostic accuracy when dealing with visual impairment, neurological and hormonal symptoms. They tend to invade locally and relapse after treatment, requiring a long follow-up.