



Sella located hemangiopericytoma mimicking non-functional pituitary adenoma

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Introduction

- Hemangiopericytoma(HPC) is a malignant vascular tumor originate from pericytes around the capillary and venules.
- Intracranial hemangiopericytomas usually arise from meninges,rarely they locate periventricular,suprasellar or sellar.
- Because of the malignancy potential,the differential diagnosis is important.
- A rare case of sellar HPC mimicking non-functional pituitary adenoma will be presented.

Case

- A 45 years old woman has admitted to our hospital complaining about two year's ongoing amenorea, spontaneous galactorea and one month ongoing headache.
- Hypocortisolism was detected at the hormonal tests and ACTH stimulation test was performed. The hypocortisolism was verified and we gave steroid therapy to the patient. The other hormonal tests were normal.
- The 3x2 cm suprasellar,well demarkated, predominantly solid with multipl cystic areas mass lesion extending into the sella was shown at the MRI images. Because of the sudden vision loss, the patient was operated as an emergency, the prediagnosis was pituitary apoplexia. Because of the bleeding, the mass couldn't been totally resected at the procedure.
- There was residual mass at the post-operative MRI images but the visual examination was improved postoperatively. Central hypothyroidism was seen after the operation so we gave the patient levotyroxine therapy. The pathological diagnosis was HPC. The patient developed acute vision after 2 months, so another surgical procedure was performed. The pathological diagnosis was HPC again.

Table-1: Laboratory test results

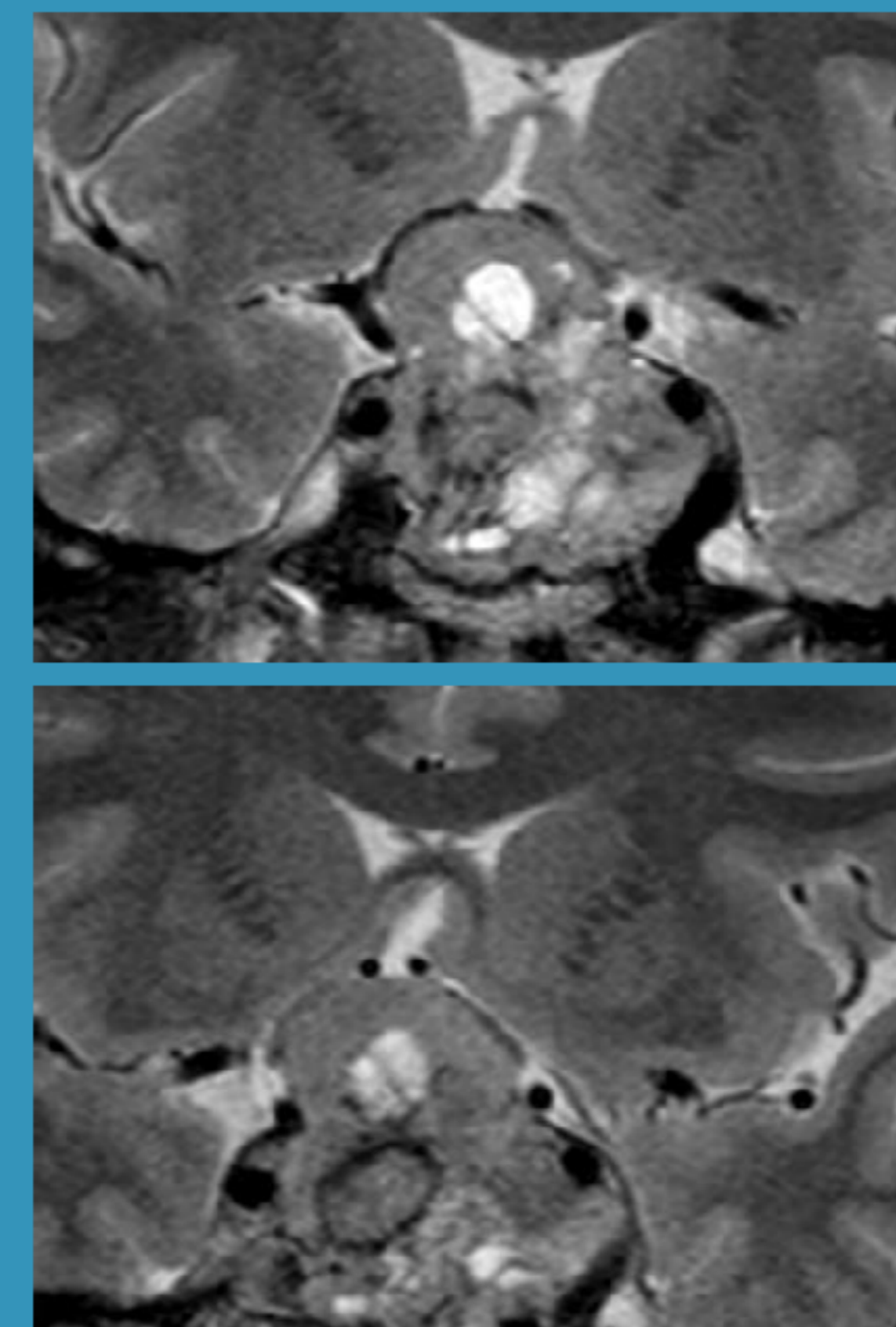
Test	Result	References
Free T4	0.54 ng/dL	0,70 – 1,48
Free T3	2.51 pg/mL	1,71 – 3,71
TSH	3.481 uIU/mL	0,350 – 4,940
FSH	4.13 mIU/mL	2,55 – 16,69
LH	0.97 mIU/mL	0,56 – 14,0
Estradiol	<10 pg/mL	21 – 312
Progesterone	<10 ng/mL	1,2 – 15,9
Prolactin	71.00 ng/mL	1,2 – 29,93
Growth Hormone	0.16 ng/mL	0,06 – 5,00
IGF-1	61 ng/mL	101 – 267
ACTH	19.7 pg/mL	5 – 46
Cortisol	1.1 ug/dL	3,7 – 19,4

TSH: thyroid stimulating hormone FSH: follicle stimulating hormone LH: luteinizing hormone
IGF-1: Insulin-like Growth factor-1 ACTH: adrenocorticotrophic hormone

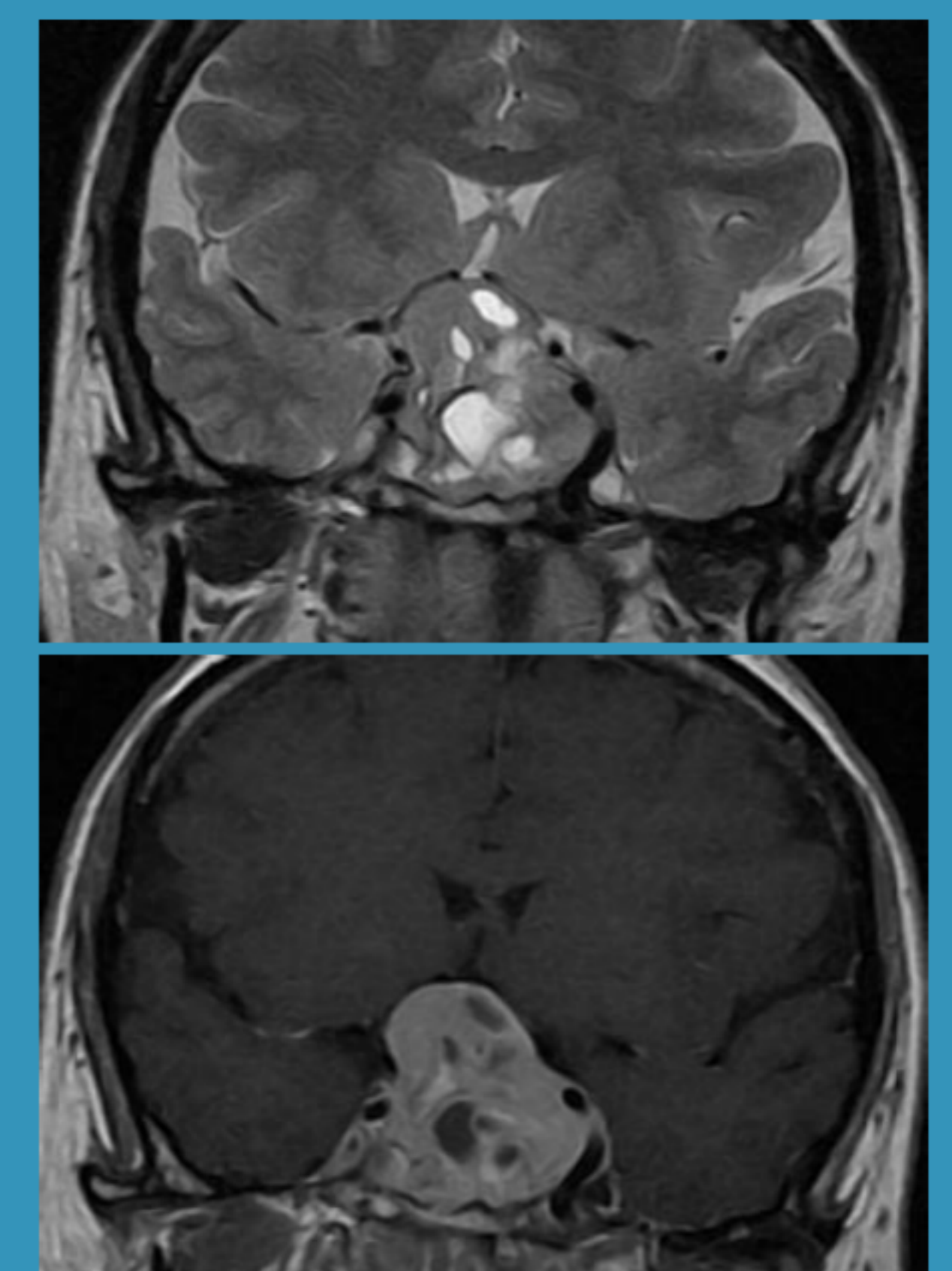
Table-2: Immunohistochemical staining results

Marker	Result	Marker	Result
EMA	-	BCL-2	+
PR	-	CD31	+
Ki 67	70/1000 positive	VIMENTIN	+
ACTH	-	DESMIN	-
PROLACTIN	-	CD99	+
FSH	-	CK	-
LH	-	S100	Focal positive
TSH	-	SMA	Focal positive
GH	-	SINAPTOFIZIN	-
P53	-	KROMOGRANIN	-
CD34	+	CD56	-

EMA: epithelial membrane antigen CD: cluster of differentiation TSH: thyroid stimulating hormone
FSH: follicle stimulating hormone LH: luteinizing hormone GH: growth hormone
ACTH: adrenocorticotrophic hormone SMA: small muscle antigen



Pre-op



Post-op

Discussion

HPC arising from central nervous system(CNS) form less than 1% of intracranial tumors and less than 2.5% of the meningeal tumors and are rarely diagnosed preoperatively. The symptoms are related with the location of the tumor. Also the duration of the symptoms are less than one year because of the rapid growth potential of the tumor. Headache and focal neurological deficits are most common at the patients with supratentorial located tumor. HPC is radiologically similar to meningioma and they can be separated from sellar adenomas by strong enhancement of gadolinium when compared with CNS and less enhancement of gadolinium when compared with hypophysis. Unlike, at HPC, a hyperdense mass with focal hypodense areas detected at CT. MRI shows the mass that has isointense enhancement of gadolinium at T1 and T2 weighted images. Dural tail sign seen at the half of the cases. "Corkscrew" vascular configuration can be seen. Still radiological diagnosis of this tumor is nearly impossible. Sellar adenomas bleed as a leakage during surgery while HPC's are vascular tumors that can not be controlled by classical bleeding control methods.

HPC relapses in many cases and metastasizes to other parts of the body. Because of the aggressivity of this tumor,it relapses locally even after total resection and metastasizes to CNS or other parts of the body. Patients that received radiotherapy(RT) have better local control, disease-free survival,and overall survival rates. In selected cases chemotherapy can be discussed at the patients who had recurrence after RT.

References

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