Case Report

Neurohypophysis Tumors.

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Abstract:-
Introduction: Neurohypophysis tumors (NHPT) named pituicytomas are very rare tumors as less than 60 cases have been reported so far. Our aim was to report two cases observed in our practice with literature review.
Observations:
Case1: A woman aged 33 consulted for polyuria and polydypsia. The diagnosis was central diabetes insipidus (CDI) without pituitary insufficiency. MRI discovered a 7mm tumor in the posterior part of the pituitary gland. According to different investigations a primary lesion was more likely than a metastasis.
Case2. A man aged 55, was referred for headaches and visual troubles. Brain MRI showed a pituitary tumor measuring 55x32mm. Hormonal exploration pleaded for a non functioning tumor with anterior pituitary deficits, without CDI. Histological examination and immunohistochemistry pleaded for a pituicytoma.
Conclusion: NHPT are deemed to be rare. They may be very small with CDI as the only manifestation as in our first case, or be large with ophthalmological troubles and pituitary insufficiency without CDI mimicking an anterior pituitary tumor as in our second case. Pituitary surgery is the best treatment for large tumors, but for small ones “wait and see attitude” can be considered as they are slow growing and much vascularized which makes surgery sometimes too risky. Radiotherapy is still debated.

Introduction
Posterior pituitary tumors (PPT) are low-grade gliomas (WHO 2007) originating from the neurohypophysis or the infundibulum\(^1\,^2\). The first case was described in 1951. These tumors are exceedingly rare. Till now less than 60 cases have been reported\(^1\). They are usually called pituicytomas as they arise from the neurohypophysis.
stroma’s cells named pituicytes. Other names are choristomas, granular cell tumors, Abrikossoff’s tumors and pilocytic astrocytoma\(^3\). They are deemed to be benign and slow growing\(^4\). Our aim was to describe two cases observed in our practice, and to review the literature about the subject.

**Observations:**

**Case 1:** a woman aged 33 consulted for polyuria and polydypsia. The diagnosis was central diabetes insipidus [(Diuresis =4.5l/24H, urine specific gravity (USG) =1005)] without pituitary insufficiency [(Cortisol =413nmol/l (n: 154 -638nmol/l); free thyroxin (FT4)=20pmol/l (n: 8-24); estradiol= 227pmol/l (n: 209-838), follicle stimulating hormone (FSH) = 8.8mu/ml (n=2-10), luteinizing hormone (LH) = 4.3mu/ml (2.2-8), growth hormone (GH)= 0.78mUI/ml (n<20), median prolactin (PRL)=37ng/ml (n=10-20)] . Cerebral MRI discovered a well limited tumor in the posterior part of the pituitary gland measuring 7 mm in height, with a lack of posterior pituitary signal (fig1).

According to our different investigations a primary lesion was more likely than breast, thyroid, pulmonary or genital metastasis. She was treated with Desmopressin which normalized urine wasting and USG. For the tumor we opted for “wait and see attitude”. After 4 years the tumour’s size is still the same.

**Case 2:** a man aged 55, consulted for headaches and decrease in visual acuity. Clinical examination showed gynecomastia (Tanner’s stage 3) with hypogonadism.
Brain MRI showed an invasive pituitary tumor measuring 55x35x32mm. The posterior pituitary signal was lacking (fig.2).

Hormonal exploration pleaded for a total pituitary insufficiency [Cortisol=44nmol/l (n: 154 -638nmol/l), FT4=3.39pmol/l (n: 8-24), Testosterone = 0.21nmol/l (n: 10-41), PRL= 4ng/ml (n=5-15), GH= 0.34mUI/ml (<20)]. Posterior pituitary function was normal: diuresis= 1.3l/24h, urine gravity=1020.

Ophthalmological examination showed a low visual acuity=1/10 for the right eye and 3/10 for the left one, and optic atrophy with degenerative maculopathy.

He was operated on, and had partial resection of the tumor without any problem. Histological examination argued for a typical pituicytoma with spindle shaped cells arranged in fascicles, with elongated nuclei (Fig.3):
Immunostaining was positive for S-100 protein, vimentin, glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA).

**Discussion**

Pituicytomas are exceedingly rare, indolent and benign tumors arising from pituicytes of the neurohypophysis and pituitary stalk. They are usually located in sellar and suprasellar region, and are mimicking non-functioning tumors as in our second observation.

In adulthood they seem more frequent in men. But, they are also observed in children, adolescent and in elderly.

Clinical symptoms are variable according to their location. Curiously, they scarcely lead to diabetes insipidus as in our second case. But, they can be revealed by visual troubles (in about 74%) or sign of infundibular compression such as hyperprolactinemia. That one may be discrete as in our case, or moderate. An association with Cushing disease is also reported. Rarely they are diagnosed incidentally, after apoplexy or discovered by autopsy.

On the histological side, pituicytomas are composed of elongated spindle-shaped cells arranged in interlacing fascicles or in a storm form structure. For immunostaining they react positively with vimentin, S-100 protein, glial fibrillary acidic protein (GFAP), and occasionally with cytoplasmic epithelial membrane antigen (EMA) as we observed it. But, they do not react with pituitary hormones, chromogranin, synaptophysin, and neurofilament protein.

These tumors raise differential diagnoses particularly with metastasis from breast, thyroid, colon, stomach, and gynecological area. When the last ones are excluded,
posterior craniopharyngioma should be excluded too. Spindle cell astrocytoma, spindle cell oncocytoma, and granular cell tumor\textsuperscript{14,15} should be discussed by the pathologist as all these tumors are entities with confusing nomenclature, histogenesis, and imaging characteristics\textsuperscript{15}. According to Phillips\textsuperscript{16} only genomic hybridation can distinguish the pituicytoma.

The best treatment for large posterior pituitary tumor is surgery. Total resection, if possible, is mandatory as subtotal resection can lead to local re-growth. Preoperative embolization can be done as the tumors are highly vascularized. Adjuvant radiotherapy is still debated as these tumors are slow growing.

For small tumor, abstention or “wait and see” attitude is still recommended as there is not any medication available for these neoformations.

Conclusion
Neurohypophysis tumors are very rare. Their aspect varies from very small tumors with diabetes insipidus to giant ones with visual troubles and pituitary deficits mimicking an anterior pituitary adenoma. Surgery is considered as the best treatment for large tumors. But, when tumor resection is incomplete, it is difficult to say if one should add radiotherapy or not. That one is still controversial because of posterior pituitary tumor low progression.

References