Common Features of Giant Prolactinoma and Paranasal Neuroendocrine Carcinoma - Case Report

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Introduction

Giant prolactinoma is a very rare tumor that due to its massive extension into surrounding structures can present more often with neurological complications such as visual defects, cranial nerve paresis or even hydrocephalus, unlike the classic prolactinoma presentation with amenorrhea, infertility and galactorrhea. On CT/MRI exams it can present as aggressive skull base tumor and its immunohistochismy (IHC) may have common features with neuroendocrine neoplasms.

Case Report

We present the case of a patient with giant prolactinoma first misdiagnosed as a paranasal sinus neuroendocrine carcinoma (PSNEC).

A 43 y.o. woman was diagnosed with a large destructive tumor of the skull base measuring 7.6 cm in the clinical context of diplopia, facial paresthesia and right retroocular pain. CT scan revealed sella turcica involvement, bilateral cavernous sinus, right sphenoid sinus and right nasal cavity extension, and compressive mass effect on the right temporal lobe and brainstem. She was known with secondary amenorrhea since 27 y.o. She underwent a biopsy and the pathology and IHC evaluation pleaded for PSNEC. IHC was diffuse positive for synaptophysin and CD 56, focally positive for chromogranin, negative for S-100 and ki 67 was positive in aprox 8% of the cells. Prolactin staining was not part of the initial IHC evaluation.

Investigations and Management

Awaiting a decision on surgery vs radiation therapy, endocrine assessment revealed a very high prolactin 34.311 ng/ml (ref 4.79-23.3 ng/ml), low gonadotropins (FSH= 6.5 uUI/ml, LH= 0.81 uUI/ml) indicating hypogonadotropic hypogonadism, a normal pituitary function in rest (TSH= 1.31 uUI/ml, ft4= 12.26 pmol/l, ACTH= 30.26 pg/ml, cortisol after stimulation with 1 mg Synachten> 60 ug/dl, IGF1= 219.2 ng/ml ref 95-251), and slightly elevated plasma CgA, NSE and 24 h urinary 5-HIA. She was also presenting bilateral galactorrhea, VI nerve paresis RE, nasal RE and nasal and temporal LE visual field impairment.

Conclusion

Evaluation of large skull base tumors must include full pituitary hormonal profile due to the giant prolactinoma’s atypical presentation and its common IHC features with neuroendocrine neoplasms.