Clinical management of pituitary teratomas and safety of rhGH replacement therapy: a Case Report

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INTRODUCTION

Teratomas comprise about 0.5% of all intracranial tumors. 15 cases of sellar teratoma have been described in the last 24 years (yrs). Teratomas occur more frequently in child. Symptoms at diagnosis are neurological defects and pan-hypopituitarism. Diagnosis can be suggested though neuroimaging (TC/RM). Neurosurgical excision is suggested as recurrence (REC) rate is extremely low in cases of complete resection. We report a case of 29-yrs female pan-hypopituitaric patient (PT) recently admitted to our Pituitary Unit.

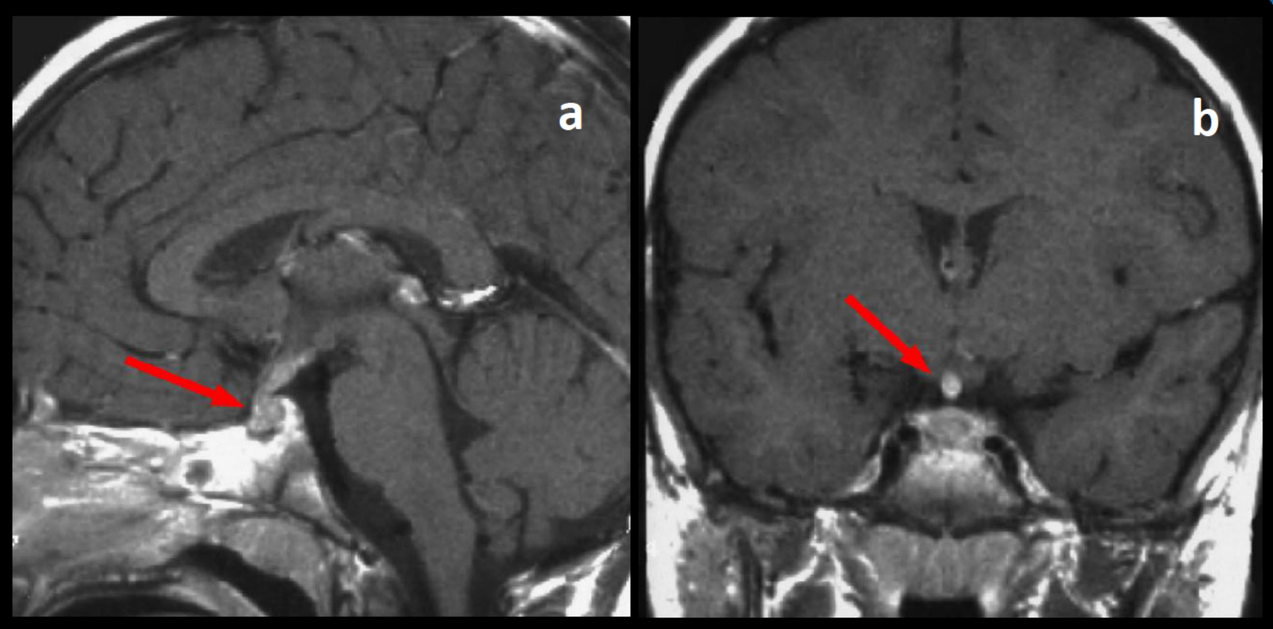

Figure 1: Coronal View (a) and sagital view (b) of RM T1 post-gadolinium with enhancement of pituitary gland and its stalk.

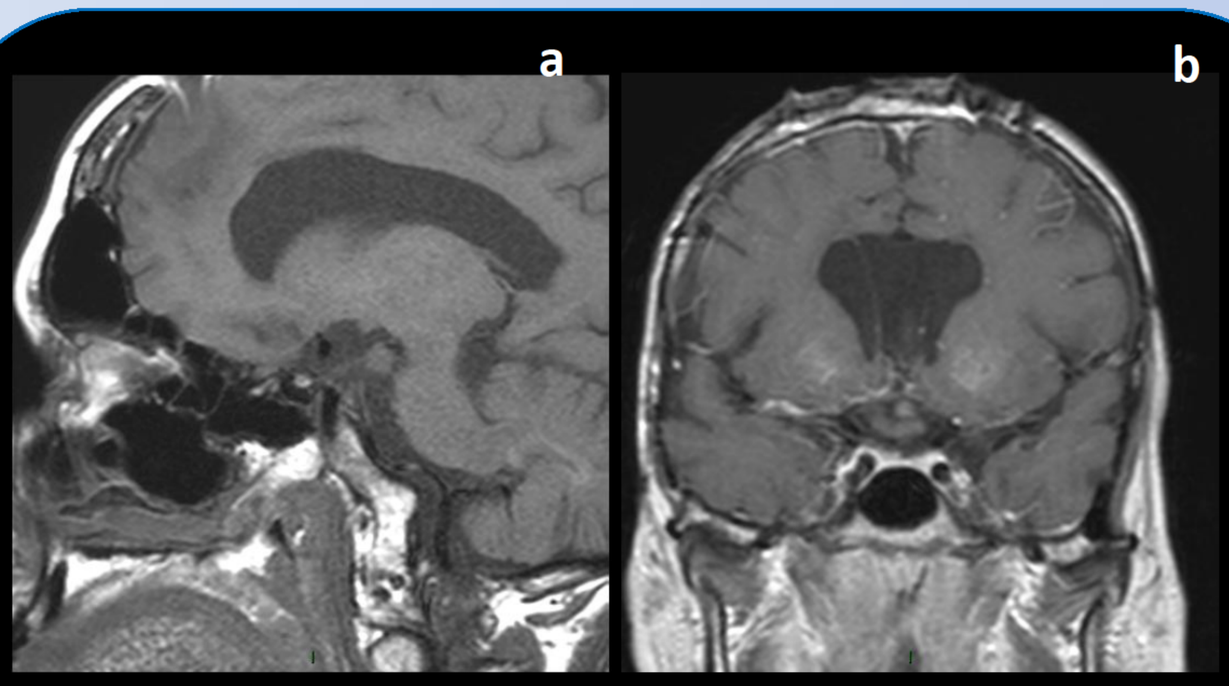

Figure 2: Coronal View (a) and sagital view (b) of RM T1 post-gadolinium with not evidence pituitary teratoma recurrence.

CASE REPORT

At 6-yrs age, for polyuria and growth delay, patient underwent endocrine evaluation which revealed diabetes insipidus and hypopituitarism. Cranial contrasted-MRI showed pituitary stalk thickening and a low intensity suprasellar lesion (Figure 1). Histological examination was suggestive for germinoma. Conventional radiotherapy (32.4Gy whole brain and 21.6 Gy to the tumor) was performed. After 6 months for disease-progression, a second biopsy was performed and histological examination was suggestive of 3th ventricle choroid plexus papilloma. Patient underwent radical neurosurgical resection. Histological examination was conclusive for mature teratoma. At 10-yrs age, patient weight was 38 kilos (Kg) (90th percentile), height was 124centimeter(cm) (3rd percentile) and growth velocity was 3cm/yr (<3rd percentile). Hormonal evaluation confirmed hypopituitarism. Clonidine test was suggestive for impaired GH secretion. Cranial contrasted-MRI didn't documented teratoma recurrence. After 3 disease-free survival years, rhGH replacement therapy (rhGH-RT) was started (somatropin 1.21mg/daily). No adverse events occurred. Every 6 months, endocrinological and neuroradiological evaluation were scheduled. At 18-years age, Cranial contrasted-MRI evidenced a falx cerebri lesion and rhGH-RT was discontinued. Patient underwent neurosurgery. Pathological diagnosis was atypical meningioma. 1-year post-surgery Cranial contrasted-MRI confirmed tumor radical resection. rhGH-RT was re-started. Every 6 months , endocrinological and neuroradiological evaluation was scheduled. At 27-years age, Cranial contrasted-MRI showed meningioma recorrence. Patient underwent second radical neurosurgery. Histological examination documented transitional meningioma with strong immunohistochemical positivity for GH receptors. rhGH-RT was discontinued. At admission in our department, PT weight and height were 100.5Kg and 170cm (BMI:34.8Kg/m2). Laboratory tests documented impaired glucose tolerance and panhypopituitarism. Cranial contrasted-MRI was negative for teratoma and meningioma recurrence (Figure 2). We suggested diet and oral hypoglycemic drugs and confirmed the rhGH-RT discontinuation.

CONCLUSIONS

To our knowledge, our case is the first describing rhGH-RT safety in a patient with teratoma history. Although rare, teratomas should be taken into account in the differential diagnosis of pituitary region tumors for allowing patients to benefit of correct treatment and later of a complete hormonal replacement therapy.

ePoster number: EP929, Pituitary-Clinical ,Chiloiro S

ECE 2016 929--EP

Pituitary - Clinical

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Poster presented at: ECE2016 28 – 31 May 2016 MUNICH, GERMANY 18th EUROPEAN CONGRESS OF ENDOCRINOLOGY