"FIRST DO NO HARM": GROWTH HORMONE (hGH) TREATMENT IN A CASE OF RECURRENT CRANIOPHARYNGIOMA.

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Introduction

- Craniohypophysialomas (CP) are benign, dysontogenetic supra or intra-sellar tumors. They are locally aggressive with severe endocrine, neurological and ophthalmological implications.[1,2]
- Lesions tend to overspread into neighboring structures and relapse after total surgical removal. Bartlett's argued that complete excision is essentially impossible his idea was supported by his postmortem examinations and high recurrence rate in almost every major surgical series.[2,3,4]
- Recurrences occur at the original site, secondary lesions (ectopic and/or metastatic) are extremely rare yet found after surgery. The possible seeding process is cell dissemination on the surgical path of passage through subarachnoid space or Virchow-Robin spaces.[1,5]

Case Report

- Patient - H.A., male, 18 years
- Medical history:
  - born naturally, late-term (42 weeks, 3200g, Apgar 7, with meconium aspiration syndrome, cared for in incubator for ~2 weeks), third child in a brotherhood of four (apparently healthy parents and brothers), slightly delayed developmental acquisitions (first steps - 1 year, talked - 2 years and 6 months);
  - OS-2011: first evaluation in Endocrinology Department (age 13 years 6 months) for growth retardation, investigations pleaded for pituitary dwarfism (short stature -2.5 SD, absence of puberty signs, delayed bone age -10 years), somatotroph axis: low basal GH=0.14ng/ml with no stimulation to arginine 30 min=0.17ng/ml, 60min=0.29ng/ml, 90min=0.27ng/ml low IGF-1=86ng/ml and rhGH therapy was initiated.
- Laboratory and imaging findings:
  - 10.2011: diagnosed with craniopharyngioma, admitted to Neurosurgery Department due to severe headache, Jacksonian seizures and optochiasmal syndrome determined by rapid development of parasellar supra/sellar invasive craniopharyngioma identified at MRI. Minimally invasive surgery was performed (partial ablation through trans-sellar approach - Fig. 1a).
- After surgery further investigations were made:
  - patient developed panhypopituitarism:
    - central hypothyroidism: low TSH levels (0.013U/ml (N<0.2-4)), low T4 levels (6.45nmol/l N<9.0-22);
    - central adrenal insufficiency: morning cortisol levels (3.4µg/dl (N=9.5-25)), ACTH levels (15.6µg/ml N<100);
    - growth hormone deficiency: low basal GH levels (0.14ng/ml), low IGF-1 (4.2mg/ml N=220-970), statural ponderal delay (height=131cm -3SD, weight=31kg), with delayed bone age-10 years;
    - pituitary-gonadal axis at prepubertary levels (Tanner I/II).
  - Post Chir.
    - GH, FSH =1.56 mIU/ml, testosterone < 0.028 ng/ml.
  - Substitutive treatment was initiated (levothyroxine and hydrocortisone with periodic dose adjustments).

Evolution

- Two other surgical procedures for tumor recurrence were performed:
  - External radiation therapy (5400/18/6/FSD).
  - Subtotal removal of the tumoral capsule.
- Further ophthalmological examination was performed:
  - Biological behavior of CPs varies considerably from case to case, some remain constant, while other lesions tend to grow over an random period of time. Subtotal removal without radiotherapy is the strongest factor in predicting CP recurrence. Radical surgical removal and the use of radiation therapy (RT) after a subtotal removal have been recognized as therapeutic practice that notably decrease the rate of CP recurrence. The presence of a high level of Ki (copious at AP exam and high KI<7 index, should be the ideal choice for close follow-up). The CP should be considered a chronic disease, requiring constant monitoring of the cases in order to provide optimal quality of life for patients, and to minimize the severe consequences of both the disease and its treatment.[10,11]
  - In addition to surgical management of CPs, substitution of endocrine deficiencies is required. CP and other central nervous system: tumor recurrence frequency is identical in hGH treated and untreated patients.[4,12]