The challenging management of aggressive, giant silent corticotroph PiTNET in a young male patient

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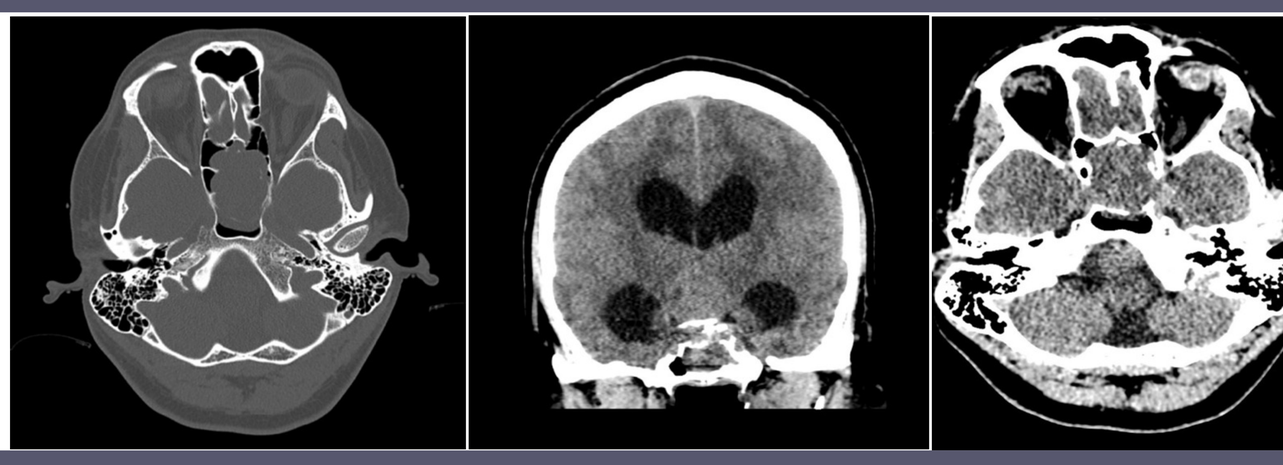
INTRODUCTION

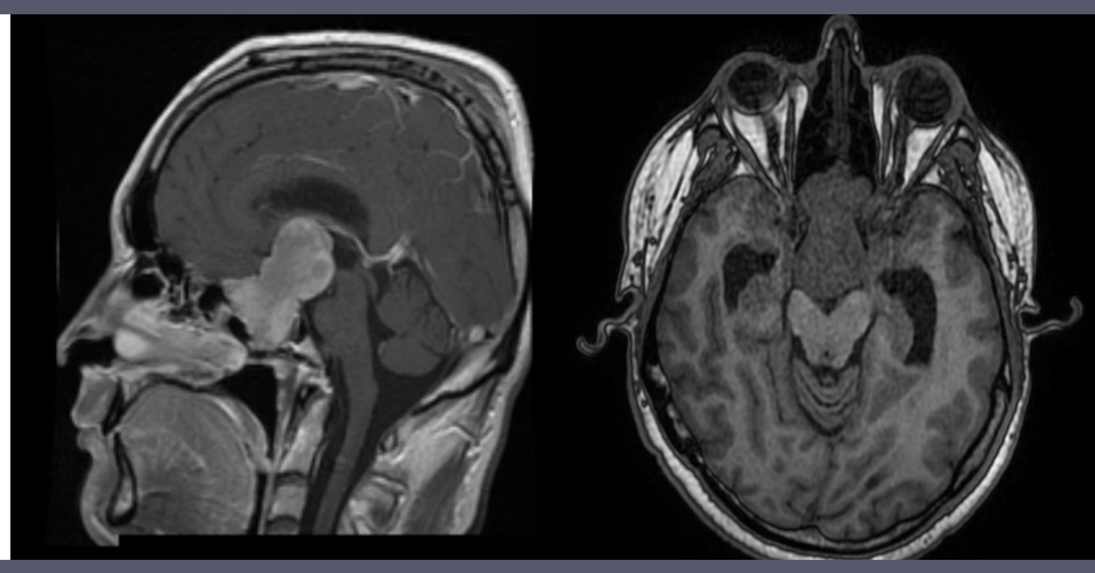
Silent corticotroph pituitary neuroendocrine tumours (PiTNET) are a subtype of nonfunctioning PiTNETs, that present positive immunostaining for adrenocorticotropin (ACTH) and/or show the expression of the transcription factor T-PIT without clinically signs of hypercortisolemia. They constitute 20% of all corticotroph tumours and manifest in most cases as macroadenoma with suprasellar extension and a higher tendency to apoplexy.

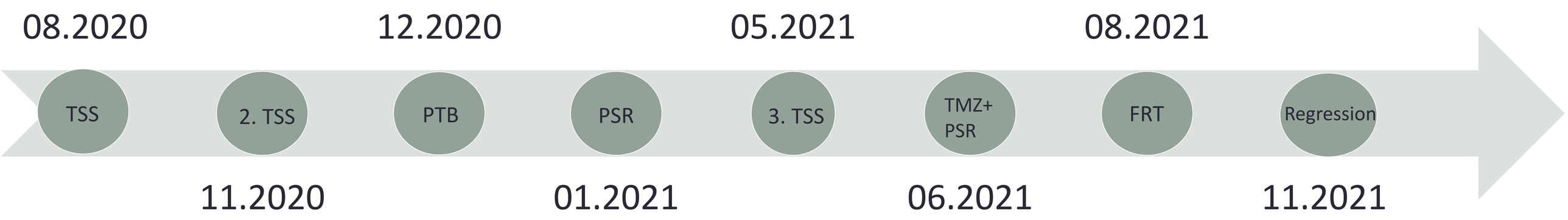
CASE

We present a 33-year-old male with aggressive course of silent corticotroph PitNET. The patient was admitted to Emergency Department due to severe headaches and vomiting. Headaches (8-9/10 using numbering rating score (NRS)) and worsening vision loss since a year. In CT a sellar tumour mass (39x33x55 mm), with extrasellar extension, causing pressure on the cerebral aqueduct of the third ventricle and cerebral edema were present.



Emergency external ventricular drainage was performed due to obstructive hydrocephalus and two days later, **debulking transsphenoidal surgery (TSS)**. **Histopathology** results showed silent adenoma subtype 1 (densely granulated), Ki67<1%. Genetic testing was negative for *AIP* and *MEN1* mutations. 3 months later, in MRI progression of PitNET was described. Subsequently, **second TSS** was performed. Biochemically, persistent multiple pituitary hormone deficiencies and diabetes insipidus were diagnosed. Clinically, severe headaches (9-10/10 using NRS) without improvement after analgesic and worsening vision loss were observed.





TSS- transphenoidal surgery; PTB- pituitary tumour board; PSR- pasireotide; TMZ- temozolomide; FRT- fractionated radiotherapy

Multidisciplinary pituitary tumour board consultation: radiotherapy was planned. **Pasireotide (10mg)** monthly and 0.5 mg of **cabergoline** weekly were scheduled. However, emergency TSS (05.2021) with the decompression of the optic nerves was performed.

After surgery, **chemotherapy with temozolomide** (starting dose of 150mg/m^2) for 5 days was introduced. After first cycles, **adjuvant stereotactic fractionated radiotherapy** (total dose 50,4 Gy in 28 cycles) was performed. Temozolomide (TMZ) at the dose of 200mg/m^2 for 5 days every 4 weeks was continued. Severe headaches (9-10/10 using NRS) without improvement after analgesic were still present. **Pasireotide (increasing dose from 10 to 40 mg/month)** was reimplemented.

Results: **Decrease of headaches from (initially 9-10 to none /10 using NRS).** In last MRI, after 5 cycles of temozolomide, and during pasireotide and cabergoline treatment, **regression of the pituitary tumour** (current measurements: 20x30x29 mm) was observed. Additionally, patients is in a very good general condition, reports no headaches.