Re-irradiation of a recurrent pineal parenchymal tumor of intermediate differentiation (PPTID): case report.

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Background and objectives

Pineal parenchymal tumor of intermediate differentiation (PPTID) was recognized in the 2007 World Health Organization (WHO) classification (1) as a new pineal parenchymal neoplasm, with intermediate malignancy (WHO grade II or III) between pineocytoma (grade I) and pineoblastoma (grade IV). PPTIDs are extremely rare entities and their optimal management has yet to be determined, especially for relapsing cases. We report a case of PPTID who was re-irradiated after leptomeningeal dissemination.

Case report

•A 51-year old man was admitted in December 2005 to our Neurology Department complaining of diplopia. We performed a *brain MRI* that showed an enhancing pineal gland mass, with restricted diffusion, infiltrating the posterior commissure, the dorsal midbrain and the antero-superior portion of the cerebellum.

•The patient underwent an **endoscopic third ventriculostomy (ETV) with tumor biopsy**; the histopathologic examination was compatible with PPTID (ki 67 8%). Therefore, we performed a *spine MRI* and a *lumbar puncture* with *cytological CSF analysis*, which excluded metastatic spinal dissemination.

•The patient then received **adjuvant conformational radiotherapy (total dose of 25 Gy in 5 fractions)** with a complete response.

•No recurrence was observed at regular follow-up until September 2011, when a *brain MRI* showed leptomeningeal spread at the anteromedial medulla, ventral left surface of the spinal cord (C2 level), knee of the corpus callosum and right superior colliculus. The patient then underwent **craniospinal irradiation (total dose of 36 Gy in 20 fractions) and sequential boosts were delivered to the new lesions (total dose of 14.4 Gy in 8 fractions**), as shown in Figure 1. After the treatment, the patient developed thrombocytopenia (grade III), bilateral optic neuropathy and ageusia, being the latter probably due to the irradiation of the solitary tract nucleus.

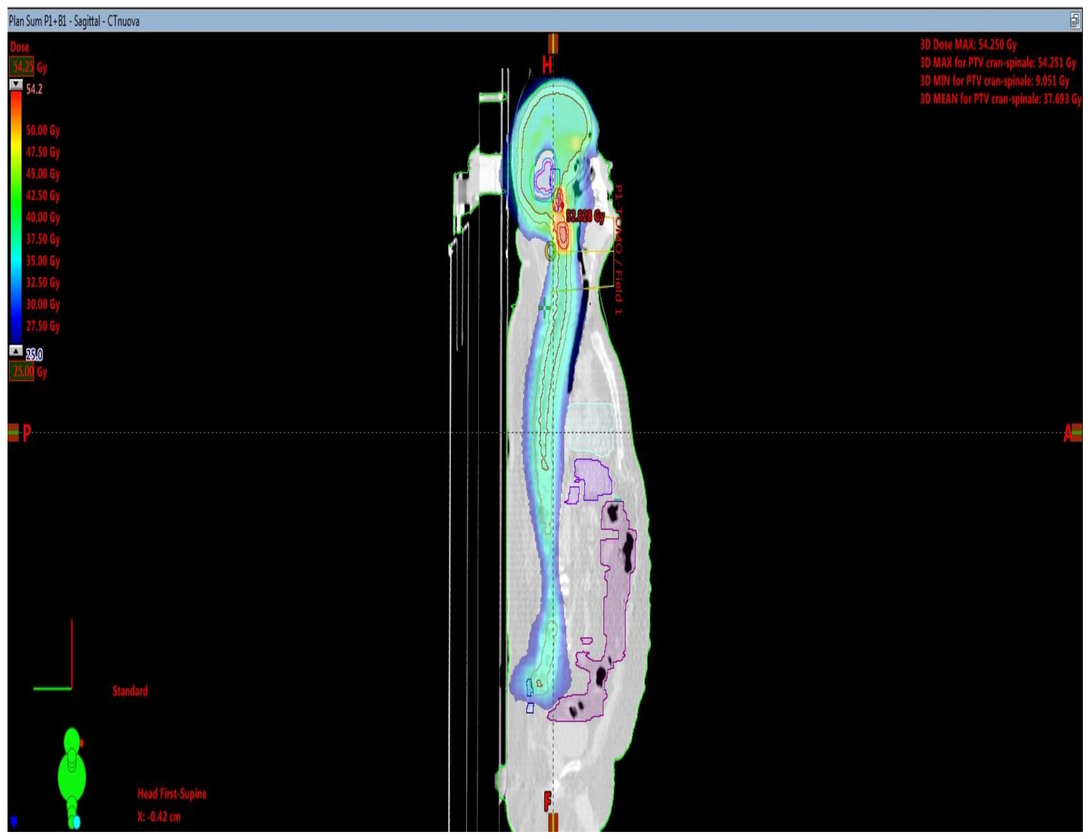

Figure 1: The radiation treatment planning after leptomeningeal dissemination of the tumor

Two years later, at regular follow-up, we observed MRI signs of radiation-induced leukoencephalopathy. No recurrence occurred until the last brain MRI (May 2016), when a new enhancing nodule appeared at cerebellar vermis.

Discussion and conclusion

Although PPTID may be aggressive and can seed through cerebrospinal fluid, PPTID patients may have a long survival, even after recurrence (2). **Radiotherapy may represent an effective therapeutic option for this type of tumor, even when a leptomeningeal dissemination has occurred**; **nevertheless, our case points out its potential complications.** Therefore, clinicians should weigh risk and benefits of this treatment for each patient.

References

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