Proton therapy as a promising therapeutic option for children with aggressive and uncontrolled pituitary macro adenoma: case report

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Background
Non functioning pituitary macro adenoma is rare during childhood. Therapeutic options are reduced to surgery and radiotherapy. Proton therapy is a particle therapy that uses a beam of protons to irradiate the tissue with the chief advantage that as a charged particle the dose is deposited over a narrow range and there is minimal exit dose.

Objective and hypotheses
Proton therapy is largely used in France for paediatric craniopharyngiomas irradiation and more recently for some adult pituitary adenomas. Proton therapy could be a good therapeutic option for benign tumor as pituitary adenoma in young child to reduce side effects observed usually with conventional radiotherapy.

Method
We report a 10 years old boy with invasive and aggressive non-functioning pituitary adenoma treated by surgery and proton therapy with more 24 months follow up.

Clinical case
Aurélien is a 10 years boy with no significant familial or personal history. He was referred for a sudden lost of visual acuity and bilateral hemianopsia. Recent decrease in growth velocity was observed but normal clinical examination (P1G1). Brain MRI showed a voluminous (3.5 cm) intra and supra sellar tumor with right cavernous sinus invasion and chiasma compression. GH and TSH deficiency were documented by endocrine testing while blood prolactin, FSH, LH, αsubunit levels were normal. The search for gene mutations were negative for AIP, NEM1 and GNAS genes.

Neurosurgeon performed partial resection first with complete visual recovery. No diabetes insipidus.

9 months after first surgery, visual defect recurrence was observed with lesion increase on MRI.

Histopathology studies confirmed: Aggressive pituitary adenoma with focal immunolabelling for FSH and αsubunit(40%), PRL<3%; P53: 3% and MIB1: 6%

Second surgery was performed with complete visual recovery and same histopathology criteria than at first. Radiotherapy was then decided : because of the young patient age and our experience for craniopharyngioma irradiation , we proposed proton therapy performed during 6 weeks (Centre de Protonthérapie, Institut Curie, Orsay, France) with 54 Gy

From the end of treatment, as long as 27 months, we didn't observe progression on tumour size on MRI with complete visual rescue and no additional endocrine deficit.

GH treatment was initiated after 1 year following with excellent catch-up growth.

Conclusion: We report the rare case of a young boy with aggressive non-functioning pituitary adenoma. Proton therapy seems a good option in replacement to conventional radiotherapy to treat it especially for children. However long-term outcome is necessary.