Controversies in management of sellar teratoid/rhabdoid (AT/RT) tumors: report of two cases mimicking pituitary adenomas and review of literature

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Controversies in management of sellar teratoid/rhabdoid (AT/RT) tumors: report of two cases mimicking pituitary adenomas and review of literature

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Background: Sellar teratoid/rhabdoid (AT/RT) tumors represent rare lesions that occur mainly in children younger than 3 years of age. Adult cases are exceptional, with only about 40 cases reported. Despite aggressive treatments, AT/RTs present poor prognosis, with a median survival of less than 1 year.
We describe the clinical, neuroradiological and histological features of two cases of adult AT/RT tumors discussing similarities and differences.
Methods: We report two cases of sellar lesions mimicking pituitary adenomas. In both cases the clinical onset was characterized by oculomotor nerves palsy. Preoperative MR study revealed sellar/suprasellar lesion invading parasellar regions (left cavernous sinuses in one case) with an intense and disomogeneous contrast enhancement. In one case, a diagnosis of micro-prolactinoma was done 10 years before, successfully treated with cabergoline. An endoscopic endonasal procedure was performed in both cases. In one case postoperative fractionated radiation therapy was started after histological examination, six weeks after surgery, whereas in the other case a high-dose chemotherapy with antraciclines was administered.
Results: A total resection of lesion was achieved in both cases. The lesion appeared grey coloured, tough and bloody. The histological examination demonstrated an atypical teratoid/rhabdoid (AT/RT) tumor.
In both patients, a slight neurological improvement was observed few days after surgery. No abnormalities of pituitary function were registered before and after surgery. A sudden clinical worsenig occurred shortly after surgery; a MR study revealed the regrowth of the sellar masses. Radiation therapy in one case, and antraciclines in the other one were started. After a transient clinical improvement and a slight volume decrease, the tumor showed a rapid progression. Both patients died 3 months after surgery.
Conclusion: We discuss the different postoperative therapeutic strategies, and speculating about the possible optimal therapeutic protocol.

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