RADIOThERAPY FOR A PINEALOBlastoma: CASE rePORT

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DOI: 10.20959/ejpmr201910.6567

ABSTRACT

Background: Pinealoblastomas (PB) are aggressive malignant brain tumors of the pineal region with challenging multimodal management. Objective: The aim of this observation is to highlight the role of a nonsurgical approach especially radiotherapy in the management of PB. The case: We report a case of a young female patient of 17 years old who presented with intracranial hypertension. The CT-scan showed a pineal mass with hydrocephalus and the MRI showed a voluminous tumor of the pineal region. The resection was not possible. A Ventriculoperitoneal shunt was made with tumor biopsy. The diagnosis of a pinealoblastoma was confirmed on pathology report. The patient received a cranio-spinal radiotherapy with a boost to the tumor and then adjuvant chemotherapy. Results: A complete remission both clinically and radiologically was observed with a follow up of 25 months. Conclusion: Radiotherapy remains a good and safe alternative for surgery in PB.

KEYWORDS: Radiotherapy, management, PB.

INTRODUCTION

Pineoblastomas are rare malignant embryonal tumors of the pineal region, accounting for <1% of all pediatric brain tumors; they are more frequently found in children and young adolescents.[1,2] On imaging and histology, PB resembles medulloblastomas and retinoblastomas. According to World Health Organization classification, it is a grade IV.[3] They are very aggressive tumors with potential cranio-spinal axis metastasis of about 15 %.[4] Symptoms result from compression of the surrounding local structures, such as the dorsal midbrain and cerebral aqueduct, resulting in ophthalmoparesis and obstructive hydrocephalus.[5] Given the rarity of PB, the management strategy is still not standardized. Surgery, radiotherapy and chemotherapy are the main treatment modalities. This case is reported to illustrate a non surgical approach for treating PB.

The case
This is a 17 years old female patient, high school student, with no prior events in her medical history.

The symptoms go back to a year ago with mild dizziness and visual blurr. The treatment was symptomatic by the general practitioner. There was no clinical improvement. After a while, there was an increase in the symptoms especially headaches and vomiting.

The patient was reffered to the university hospital Hassan II for management.

After admission, a clinical examination showed a patient who was conscious, with nausea and vomits. An ophthalmological examination showed a decrease in visual acuity in both side a divergent strabismus with no papillary edema. There was no neurological deficit. It was a clinical stage of intracranial hypertension with no signs of infection.

A brain CT-scan was made and showed: a pineal tumor with triventricular active hydrocephalus (Figure 1).

A brain MRI showed: Bulky lesional process centered on the pineal region with lobulated contours, with a double fleshy component, and cystic majority, with some calcifications, without hemorrhagic stigmas. It is in isosignal in T1 weighted sequence, with an intermediate signal in restrictive T2 in diffusion, and which is enhanced moderately, and progressive after contrast (gadolinium). This process measures 58x32x40 mm. It invades the splenium of the corpus callosum, compresses the V3, the Sylvius aqueduct, the V4, and pushes the mesencephalon forward and the vermis downwards thus responsible for a descent of the cerebellar tonsils to 3mm above the line of Mac Rae, and symmetrical triventricular dilatation (ventricular junctions measured at 20mm, and a V3 measuring 12.5mm maximum
transverse diameter), with signs of trans-ependymal resorption. Dilatation of peri-nerval sheaths of optic nerves in relation to HITC signs (Figure 2).

The diagnosis that were evoked at this stage were: germ cell tumor or malignant pineal tumor.

The neurosurgery team performed a ventriculo-peritoneal shunt to relieve the hydrocephalus symptoms. After discussion at the tumor board, they decided to perform a stereotactic biopsy given the fact that a resection would be at high risk with the critical structures close to the tumor.

The pathology report of the biopsy stated: blastematous tumor formation of high cell density, slightly differentiated cells with a little or no visible cytoplasm. The nuclei are ovoid, hyperchromatic with little or no nucleolate. Anisokaryosis is marked. Here was no seen cell arrangement. the whole is partitioned by fibrous strips of variable thickness. Immunohistochemistry: anti-neurofilament antibodies positive, anti-synaptophysin antibodies positive. Ki-67: 70%. They concluded to a pinealoblastoma.

A cerebro-spinal MRI then was performed showing no metastasis at this level at any level.

After tumor board discussion, it was decided to treat with radiotherapy and if needed adjuvant chemotherapy, resection being risky and unfeasible.

The patient was reffered to our department for management.

The protocol was a cranio-spinal radiotherapy of 36 Gy and then a boost to the tumor to a total dose of 54 Gy. The fractionation was 1.8 per fraction.

A simulation CT was performed C- and C+ in a prone position with a head thermoplastic mask for immobilization.

The target volumes were countoured after MRI-CT fusion: GTV (gross tumor volume), CTV (clinical target volume : CTV=GTV + 0.5 cm) and PTV (planning target volume : PTV=CTV+ 0.5 cm).

The radiation technique used was 3D conformal radiotherapy with lateral fields for the brain and the boost and anterior fields for the spine.

The treatment tolerance was good. There was a decrease in symptoms throughout the radiotherapy course.

First control MRI after three months: Presence of a tumor residue in isosignal T1 and T2 with double component fleshy and cystic with some area in signal T2, enhanced after gadolinium measuring 15x20x10 mm. Total regression of hydrocephalus (Figure 3).

After this result, the medical oncology team decide to add adjuvant chemotherapy with etoposide -cisplatin every 21 days for a total of 6 cycles.

A pre-chemotherapy assessment was performed with blood formula, renal and hepatic function, cardiovascular evaluation.

The chemotherapy protocol: Cisplatin 75mg/m2 day 1- Etoposide 100mg/j day 1 to day 3 with G-CSF. Day1=Day21.

The tolerance was moderate and a total of 6 cycles were completed.

Another control MRI was performed one year later: Persistence of a small residue on the opto-chiasmatic tank of 4 mm extended on 13 mm versus 17 mm (Figure 4).

The clinical examination showed a complete remission in term of headaches, visual blur and visual acuity. The patient actually is living a normal life.

Her last MRI (25 months after radiotherapy) showed a complete remission with no remaining tumor (Figure 5).

The patient is still doing well and is on follow-up every 6 months.

Figures
DISCUSSION
Pinealoblastoma are primitive embryonal malignant tumors.\(^6\) Compared to other pineal region tumors, pineoblastomas typically have greater cytologic atypia, more frequent mitotic activity, and lower relative expression of neuronal markers.\(^7\) They are similar to PNET and sometimes described as supratentorial PNET. Radiologically, pinealoblastomas contain calcifications that spread throughout the lesion (most seen on CT). On MRI, in diffusion sequences, there is a restricted
diffusion. Multiple components (fleshy and cystic) is not characteristic. In addition, there a more tendency to hydrocephalus in PB. Some studies reported that GTR may play a great role in the treatment of PB.[9] The problem remains the perioperative risk given the localization next to critical structures. Surgical mortality can go up to 7% and 10% respectively.[9] Radiotherapy is a crucial factor impacting patient outcome, but its association with long-term morbidity, particularly in young children.[10] Typical radiotherapy protocols for pineoblastomas are 55 Gy to the tumor region and 36 Gy to the spinal axis.[11] There are more studies showing a significant role of stereotactic radiosurgery.[12] Concerning chemotherapy, PB seems to be chemosensitive but without radiation therapy it does not prolonge survival.[13] Its use remains controversial. Some series reported partial response[14], while others showed no advantage to chemotherapy in terms of survival.[15] The survival is poor. Prognosis factors include in some series: younger age and presence of metastasis.[16] Cranial irradiation > or =40 Gy (p=0.014) and gross total resection (p=0.034) were associated with improved survival. There was a trend towards improved survival for women (p=0.099).[17]

CONCLUSION
Pineoblastoma tumors are rare, predominantly pediatric, high-grade tumors. Complete surgery if possible is preferable. Adjuvant radiotherapy is beneficial. Chemotherapy is controversial. When surgery is not possible, radiotherapy, especially with recent techniques, is a good alternative. There is a need for multi-institutional studies for best standardizing and understanding of treatment strategies.

REFERENCES