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Rare Case of Pinealoblastoma in Adults:

Diagnostic and Therapeutic Aspect in a Low-Resource Country

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Abstract

Adult pinealoblastoma is a rare tumor of the central nervous system. Little is known about the clinical features and outcomes of this condition in adults. The optimal therapeutic strategy for pinealoblastoma in adults remains to be determined. This case describes a 31-year-old adult with pinealoblastoma. Given the potential impact on quality of life and the technical platform, the patient did not undergo tumor resection. The patient was treated with exclusive radiotherapy, followed by chemotherapy. The patient was without recurrence in the 36 months following radiotherapy. This case highlights a minimally invasive strategy for the treatment of rare pineal region tumors in close contact with critical structures, resulting in favorable responses and excellent neurological outcomes. The radiographic and histopathological features of pinealoblastomas are also reviewed, and the various treatment options reported in the literature are discussed.

Keywords: *Pinealoblastoma, Brain Tumor.*

Introduction

Pinealoblastoma is a malignant and rare type of supratentorial primitive neuroectodermal tumor **(**PNET**)**. It is often found in children, less than 10% of cases are reported in adults. It is generally associated with a poor prognosis because it is the most aggressive pineal parenchymal tumor **(**1**)**.

We report a rare case of pinealoblastoma occurring in a 31-year-old young adult treated in our department. This case highlights the complexity of management and the need for rapid histological diagnosis. The place of radiotherapy is also important in view of the supratentorial location of the tumor, which makes the surgical approach difficult.

Observation

Mr AEB, 31, is a young man with no significant personal or family history. The onset of symptoms dates back to about 6 months with balance and gait disorders with a decrease in visual acuity and strabismus of the left eye. These symptoms motivate consultation with a neurosurgeon in private where the initial

physical examination finds a patient WHO 2. On neurological examination we find a conscious patient with a Glasgow score of 15. He is hemodynamically and respiratory stable with blood pressure at 130/80 mmHg. There is also a widening of the support polygon as well as a drunken gait. Romberg's sign is present on the right. Hypermetria to the heel-knee maneuver with paralysis of gaze elevation and vertical nystagmus; no motor sensory deficit; the osteo tendon reflexes are present and symmetrical. An optical coherence tomography carried out concluded at the level of the left eye with a temporo-macular serous retinal detachment respecting the retro-foveolar part **(**Figure 1**)**. The imaging assessment made of magnetic resonance imaging highlights an intra-ventricular expansive process of approximately 3cm x 2.9cm sitting at the level of the 3rd ventricle extended to the aqueduct of Sylvius of tissue signal compatible with an ependymoma . The process comes into contact with the midbrain and the cerebellar vermis without signs of invasion; it is responsible for tri-ventricular hydrocephalus affecting the 3rd ventricle and the VLs without signs of transependymal resorption. At the level of the cervical spine, the cervical cord is of normal caliber and of homogeneous signal **(**Figure 2**)**.

He then benefited from a ventriculo-cisternostomy between the floor of V3 and the interpeduncular cistern and a stereotactic biopsy. Examination of the biopsy specimen notes the presence of a tumor-like focus with round cells whose immunohystochemical study finds positivity for synaptophysin and CD56 on tumor cells without expression of PLAP in the presence of a positive external control. The CD 117 marker is expressed more markedly in tumor areas, however, in the absence of PLAP expression and given the expression of neuroendocrine markers, the diagnosis of a germ cell tumor cannot be confirmed. The immunohistochemical aspect is rather in favor of a pinealoblastoma. MRI-based differential diagnoses included germ cell tumor, glial tumor, or ependymoma. The biological assessments made of the dosage of tumor markers namely beta HCG, alpha feto-protein, carcinoembryonic antigen as well as the analysis of the cerebrospinal fluid all returned to normal.

The patient's file is then discussed in a multidisciplinary neuro-oncological consultation meeting and the decision was to perform exclusive radiotherapy.

The patient then benefits from a centering scan in a thin millimeter section going from the vertex to the union of the upper third, lower two thirds of the thighs.

The organs at risk as well as the target volumes were delineated after fusion-registration with magnetic resonance imaging. A total dose of 36Gy was delivered to the craniospinal axis in 18 daily fractions of

2Gy. An additional dose was applied to the tumor at 54Gy, still in fractionation of 2 Gy. The ballistics required 6 Mev photon beams using an intensity modulation technique **(**Figure 3**)**.

The total spread of the treatment was thirty-eight days; we did not find any interruption of treatment of more than two days. No toxicity of grade greater than two was found in our patient during treatment.

A post-therapeutic evaluation by MRI 2 months after the end of irradiation found an almost complete tumor response.

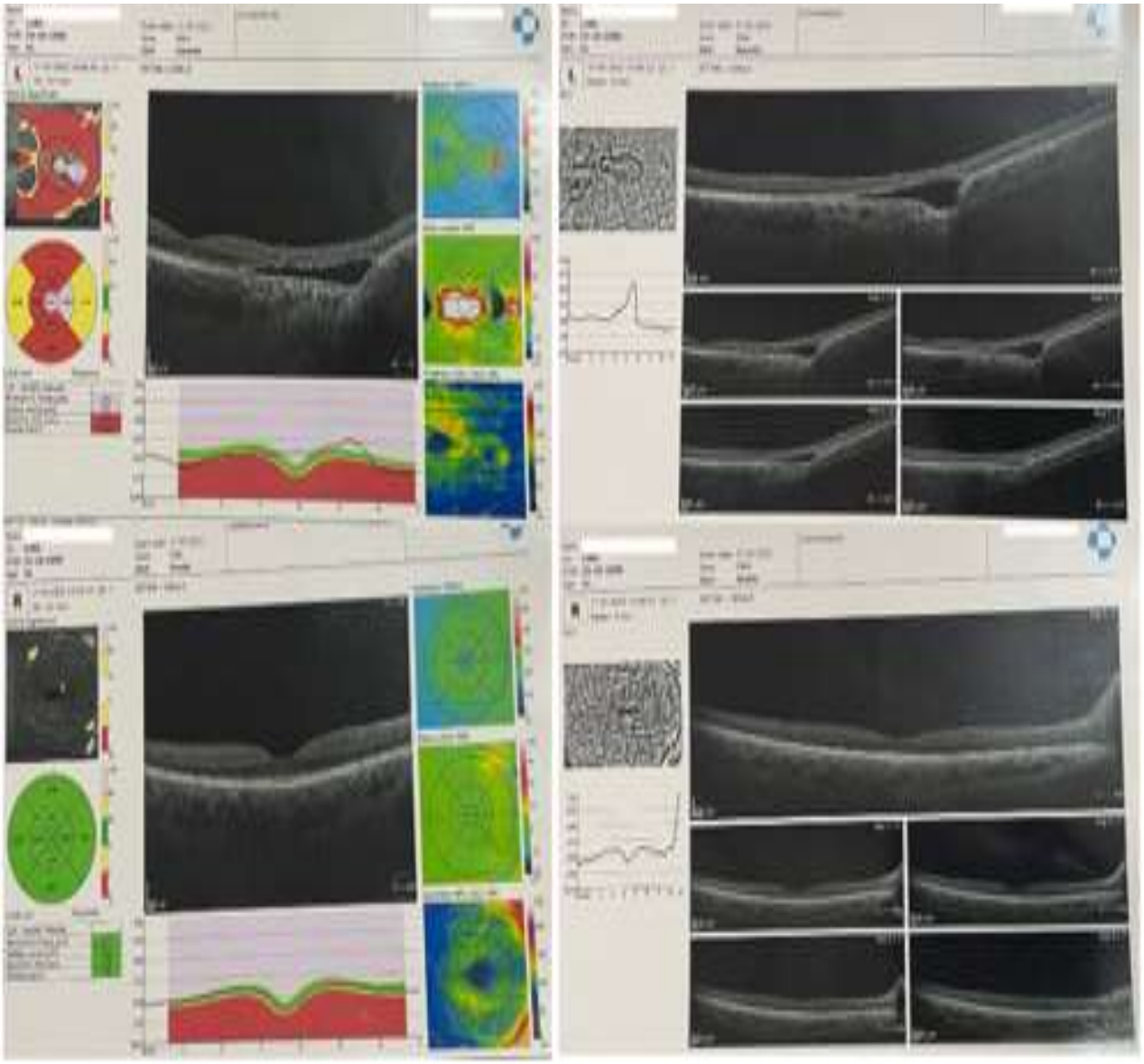

Figure 1: Optical coherence tomography of our patient showing in the left eye a temporo-macular serous retinal detachment respecting the retro-foveolar part

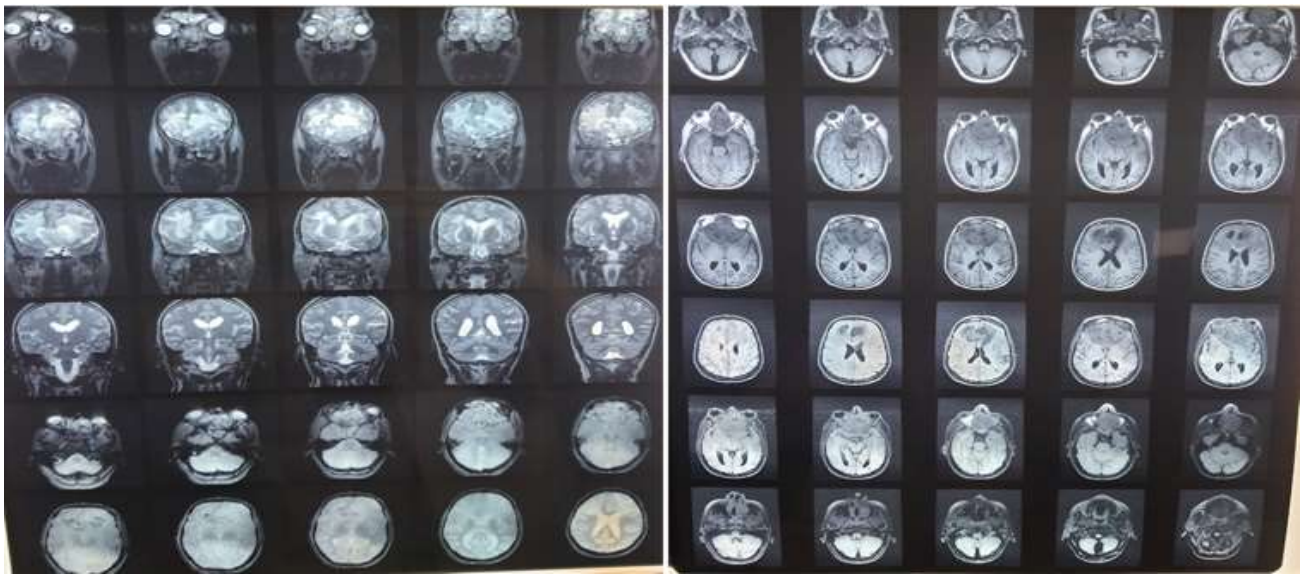

Figure 2: scannographic image of the intracerebral process of our patient

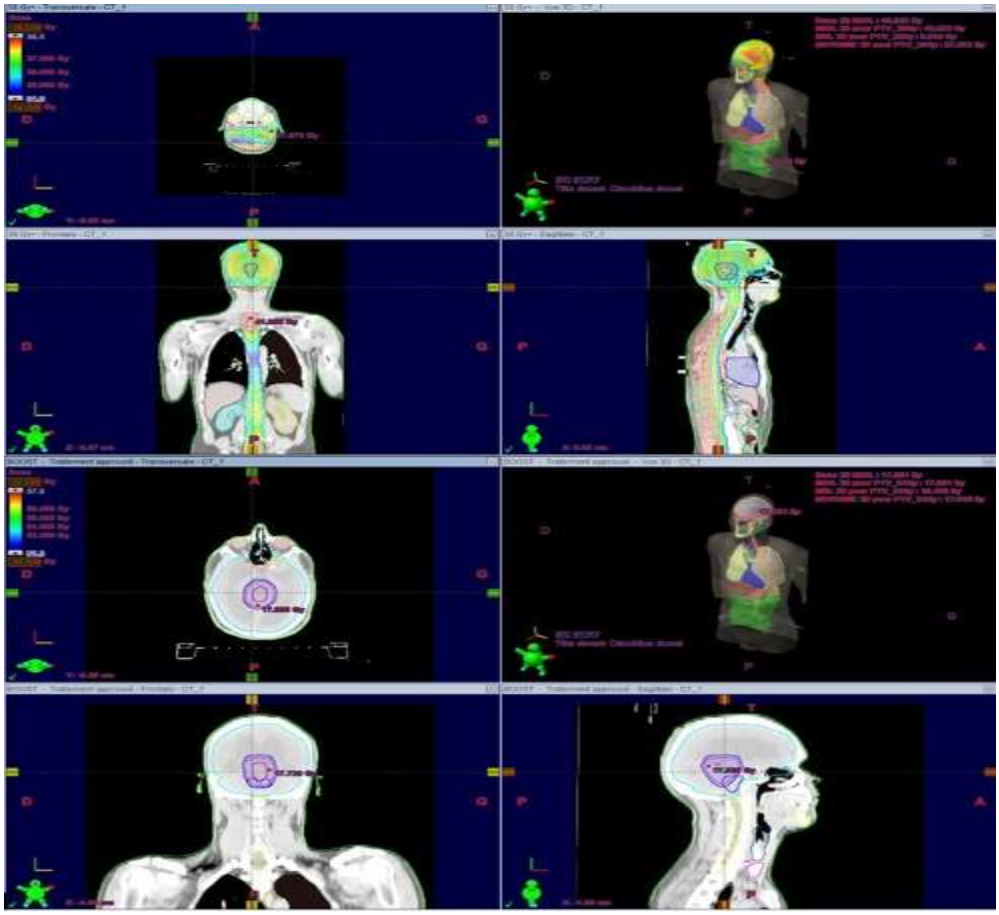

Figure3: Dosimetics images of cranio-spinal irradiation of our patient in 3D-CRT

Discussion

Adult pinealoblastoma is an extremely rare tumor of the central nervous system, categorized as a supratentorial primitive neuroectodermal tumor (PNET) localized in the pineal gland, with a tendency to regional dissemination along the neuraxis and local recurrence(2) . Despite general similarities, the morphological and immunohistochemical features of pinealoblastomas differ from those of infratentorial PNETs (e.g. medulloblastomas). We find in the literature a more unfavorable prognosis of pinealoblastoma in adults compared to children(3). There are very few data in the literature concerning the clinical characteristics and management of pinealoblastoma in adults. Among the published cases of pinealoblastoma, there is no consensus on the results between CT and MRI. On CT, the tumor tends to be large, lobulated and heterogeneously enhanced with rare calcifications(4). In addition, patients with pinealoblastoma generally presented with more marked hydrocephalus than pineocytomas. The tumors are usually solid and sometimes cystic, although this is more common in pineocytomas (5). On MRI, pinealoblastoma is generally characterized by hyposignal or hypo to iso signal on T1-weighted images, iso signal or iso to hypersignal on T2-weighted images and heterogeneous enhancement of the pineal region (6). Our patient presented several of his radiological characteristics. Histologically, pinealoblastoma does not differ much from other PNETs, in fact it has been described as a supratentorial PNET (2). PB is a highly cellular tumor composed of small, round, poorly differentiated cells arranged in sheets or unstructured aggregates. The cells contain hyperchromatic round or oval nuclei and sparse cytoplasm and are usually arranged in Homer-Wright rosettes, widely considered to represent failed attempts at neuroblastic differentiation (2). Mitosis is often seen with rosettes and areas of necrosis(7). In addition, tumor cells typically exhibit immunoreactivity for neuronal markers, such as neurofilament, synaptophysin, chromogranin A, glial fibrillar protein, and S-100 protein. In our case, the majority of tumor cells exhibited synaptophysin immunoreactivity. The pathological diagnosis of pinealoblastoma mainly depends on the location and morphology of the lesion. Pinealoblastomas should be differentiated from other types of tumors located in the pineal region, including pineocytomas, germ cell tumors, and glial tumors.

No clear treatment strategy appears in the literature because the incidence rate of the disease is extremely low, especially in adults, and only a few cases are described with a median follow-up and very limited results (8–12) . Previous studies have reported that macroscopic tumor resection may play a major role in the treatment of pinealoblastoma (13,14). However, despite improvements in surgical techniques and resuscitation, surgery of the pineal region remains cumbersome and correlated with surgical mortality

rates of around 4-7% and that of morbid sequelae can reach 10% (15) . In the case of our patient, the tumor was not amenable to complete surgery due to its proximity to the midbrain and thalamus, and intraventricular expansion. Considering the quality of life, a non-invasive radiotherapy strategy was adopted which contributed to a favorable response with complete regression of the tumor and excellent neurological results.

The effectiveness of postoperative therapy remains undefined in the literature. A few cases described reported that radiation therapy helped control the tumor and improve survival in patients with pinealoblastoma (16,17). However, these benefits have not been assessed statistically due to small sample size and lack of uniformity in radiobiological strategies and doses. Lee et al (14) examined treatment factors that influenced survival in 34 adult patients who presented with pinealoblastoma between 1969 and 1998, and found that the median survival of patients who received cranial irradiation ≥40 Gy was three times higher than that of patients who received lower doses (29.8 versus 8.1 months). However, no prospective study has to date confirmed the effect of radiotherapy or the optimal radiobiological doses in pinealoblastomas.

The use of chemotherapy remains controversial in cases of pinealoblastoma. Hinkes et al (17) demonstrated partial responses to chemotherapy in their series of six patients with pinealoblastoma. However, Lee et al (14) reported that chemotherapy conferred no survival benefit in a series of 34 adult patients with pinealoblastoma, 10 of whom received chemotherapy. None of these two studies, however, can answer the question of the benefit of chemotherapy.

In the present case, the volume of the tumor was large and its location was close to vital structures; therefore, a definitive radiotherapy was decided in order to best preserve the quality of life. Given the tendency of pinealoblastoma to disseminate into the cerebrospinal fluid, prophylactic craniospinal irradiation was administered at a dose of 36 Gy. In addition, this tumor is less sensitive to radiotherapy than germinoma and medulloblastoma (18,19), and tumor volume was large, a local "boost" at the tumor site of 18 Gy in 2 Gy fractions was administered. Adjuvant chemotherapy was also administered. The treatment allowed an almost complete regression of the tumor without neurological deficits.

Conclusion

Pinealoblastoma is rare in adults and, although an appropriate treatment for this pathology remains to be determined, the present case successfully demonstrates that aggressive surgery can be avoided in patients whose tumors show proximity to critical structures. . Taking into account the surgical complications, radiotherapy remains a feasible option as an optimal non-invasive treatment strategy. However, prospective studies including larger patient populations are needed to demonstrate the efficacy of chemotherapy and radiotherapy dose to establish a true standard for optimal management of pinealoblastoma in adults.

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