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ANAPLASTIC ASTROCYTOMA OF THE PINEAL REGION, 4-YEAR FOLLOW-UP, CASE REPORT.

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Abstract

Introduction: The pineal region tumors represent a heterogeneous variety of primary neoplasms in the central nervous system. This type of tumor has a low frequency in this region and the prognosis for survival in this type of tumor is relatively short.

Method: We present the case of a 16-year-old female with the histologic diagnosis of anaplastic astrocytoma from the pineal region with the diagnosis, treatment, follow-up, and survival for 4 years.

Discussion: According to our review, anaplastic astrocytoma has a short survival, and the parenchymal pineal tumors are less frequent than germinomatous tumors. The treatment chosen in this case was based on the international recommendations obtaining a 4-year follow-up and survival that make this case a success, contrary to the common prognosis of its histology.

Conclusion: The pineal region anaplastic astrocytoma is a tumor with low frequency. Probably, undertaking a full surgical resection, altogether with the corresponding medical handling, had a great impact on the fact that the patient had a disease-free survival above the average. We hope to contribute a case to the series of this rare tumor with a disease-free and stable evolution.

Keywords: *Alpha-fetoprotein (AFP), beta-human chorionic gonadotropin (HCG-b), Cerebrospinal fluid (CSF), magnetic resonance imaging (MRI), Isocitrate dehydrogenase (IDH), World Health Organization (WHO).*

Background

The pineal gland is a small endocrine structure, located in the epithalamus, in the cerebral midline, just rostral and posterior to the roof of the third ventricle [1]. Among some of its functions, it is involved in gonadal function and circadian rhythm [2,3]. 95% of the glandular cells are pinealocytes with dendritic processes and the other 5% are neuroglial cells including astrocytes [3].

Brain tumors are the most frequent solid neoplasms in childhood and the leading cause of cancer death[4]. Pineal region tumors account for about 0.4 to 1% of all intracranial neoplasms in the general population, including a wide variety of germ cell tumors, pinealocytomas, metastases, and extrapineal tumors dependent on surrounding parenchyma[4]. In the pediatric population, they account for approximately 3-11% of brain tumors[5].

Gliomas in this region are extremely rare, representing 14-22% of pineal tumors[5]. They were classified into diffuse and non-diffuse, using molecular features including IDH mutation. They can originate from the pineal tissue itself or the thalamus[6]. Astrocytomas grow from astrocytes of the pineal gland itself. Grade II astrocytomas are the most frequent with 25%.

High-grade astrocytomas (anaplastic and glioblastoma) are very rare in this region[4,7]. Clear differences in survival between low and high-grade lesions have been reported[2,8].

Anaplastic astrocytoma is a malignant, infiltrative, and diffuse primary tumor of the central nervous system. The management of anaplastic astrocytoma includes maximal safe resection, followed by chemotherapy and radiotherapy (the latter in patients older than 3 years)[6]. Immunotherapy represents a promising tool in the treatment of high-grade gliomas[2,6]. The surgical approach to the pineal region is chosen specifically based on the location of the lesion and the surgeon's preference[7,8].

Histologically, anaplastic astrocytoma presents a heterogeneous morphology, with the following characteristics: high cellularity, high percentage of mitoses, pleomorphism, nuclear atypia, presence of glial markers, absence of neuronal markers, and finally absence of necrosis and cell proliferation.

Tumor recurrence and progression are very frequent and therapeutic alternatives are very limited. A second intervention can be considered if the patient presents symptoms due to mass effect, when progression is not found in eloquent areas and the patient presents good clinical conditions. Surgery presents good results for symptom relief but is not very significant in the impact on survival. Radiotherapy after surgery is the established adjuvant treatment for malignant gliomas, showing a favorable clinical response. The median survival of anaplastic astrocytoma has been described to be between 21 to 25 months and the median disease-free progression is between 8.7 and 14.8 months.

Case presentation.

A 16-year-old female patient presented with headache, vomiting, and generalized tonic-clonic seizures in June 2019. An MRI study was performed (Figure 1 A.) where data of hydrocephalus and lesion in the pineal region were observed, so a ventriculoperitoneal shunt was performed, and tumor markers were taken in cerebrospinal fluid, which were negative. In August, a biopsy guided by stereotaxy was taken, which reported anaplastic astrocytoma (Figure 2.A.B.C). In September, the patient underwent resection through an infratentorial supracerebellar approach, performing a total resection (Figure 1.B). Subsequently, she underwent 6 cycles of chemotherapy with ICE-T scheme (ifosfamide, carboplatin, etoposide with temozolamide) and radiotherapy with 52 Gy in 29 sessions. Subsequently, a control imaging study was performed in 2020, where recurrence vs. radionecrosis was suspected, concluding in the latter. It was decided to offer a new chemotherapy scheme, bevacizumab and irinotecan with subsequent follow-up. In the last imaging study performed at the end of 2022, the patient was free of recurrence (Figure 1.C).

Discussion

In the case presented, there were diagnostic suspicions when observing the initial imaging study, as well as the negative cerebrospinal fluid tumor markers, so the following part of the protocol of the diagnostic and therapeutic approach was proposed.

Pineal region tumors are rare intracranial neoplasms. High-grade gliomas such as anaplastic astrocytoma and glioblastoma multiforme have a poor prognosis[4]. A recent publication by Li et al. compared the survival of low-grade (WHO Grade I and II) and high-grade (WHO Grade III and IV) gliomas in the pineal region. The median survival was 46% for low-grade at 24 months in contrast to high-grade less than 16% at 23 months[6].

Surgical treatment in some series [6] does not report a significant difference in survival, with survival being no longer than 23 months in patients with a high-grade glioma. In our case, the patient currently has a greater survival than this, where we consider that surgical resection was of great importance in obtaining these results.

Of the approaches described for the surgical resection of pineal region tumors[12], the infratentorial supracerebellar approach used in this case provided us with a good surgical corridor to offer a total resection, without subsequent added deficit. The use of bevacizumab as second-line therapy in high-grade glioma recurrences shows an increase in survival[15]. The fact of having performed a total resection and the concomitant use of bevacizumab in the case presented is in agreement with what is described in the international literature.

Conclusion

Anaplastic astrocytoma of the pineal region is a rare tumor with limited survival. Probably having performed a complete surgical resection, together with adjuvant treatment, had a great impact on the patient's survival, which was above

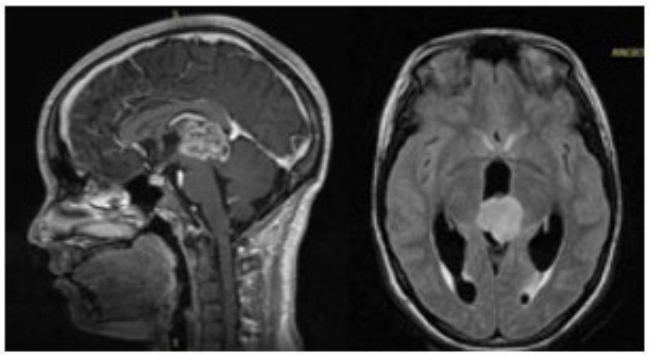

Figure 1 A

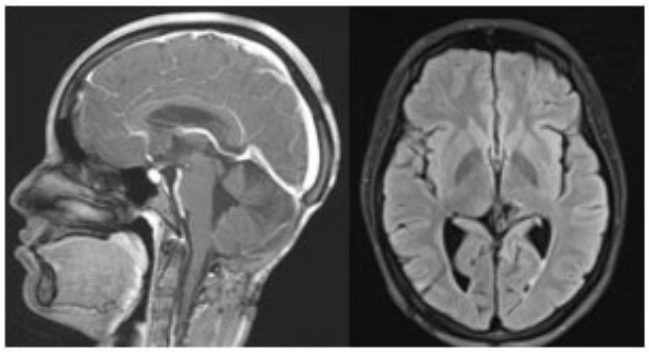

Figure1 B

Preoperative magnetic resonance imaging of the brain shows an image compatible with a tumor in the pineal region (Figure 1.A). Last MRI study of December 2022 where there is no evidence of tumors (Figure 1.C).

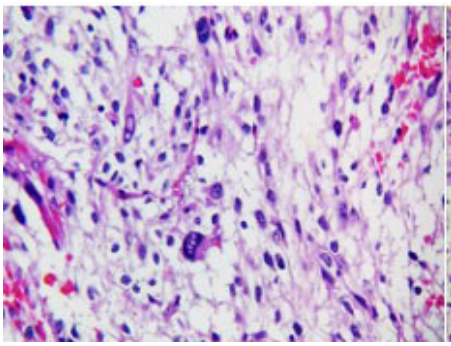

Figure 2 A. Hypercellularity with highly anaplastic glial cells with nuclear atypia and significant pleomorphism, as well as atypical mitoses (H&E stain, 40x).

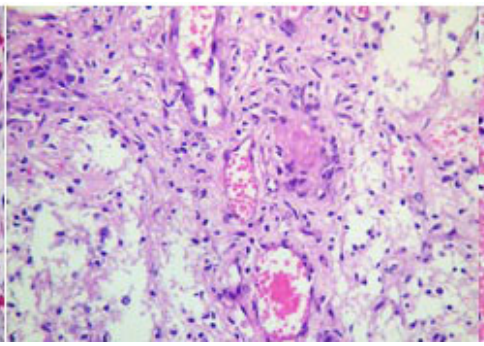

Figure 2 B. Microvascular proliferation with endothelial hyperplasia: "glomeruloid vessels". (H&E stain, 40x).

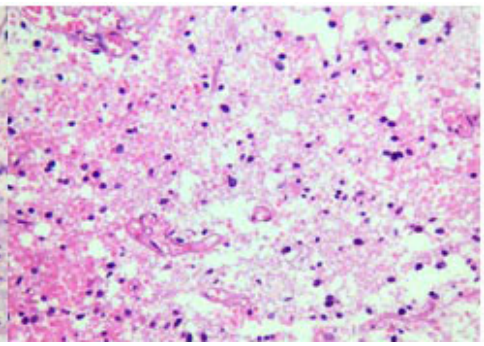

Figure 2 C. Patchy necrosis (without peripheral palisading). An essential feature in high-grade gliomas (H&E stain, 40x).

average. By presenting this case, we intend to contribute a new report of disease-free evolution to the series, in the future and with the application in our environment of the 2021 classification proposed by Cohen[14].

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Metasearch engines. PubMed.

Conflicts of interest

The authors of this manuscript have no conflicts of interest to declare.

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