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Pineal Parenchymal Tumor of Intermediate Differentiation: A Case Report and Literature Review

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Abstract **Case Report**

Background: Pineal parenchymal tumor of intermediate differentiation [PPTID] is a rare central nervous system neoplasm representing an intermediate entity between pineocytoma and pineoblastoma. Due to its low incidence and heterogeneous behavior, optimal management strategies remain controversial, particularly regarding the role of adjuvant therapies. ***Case presentation:*** We report the case of a 38-year-old previously healthy woman who presented with rapidly progressive confusion and headaches, followed by aphasia. Brain magnetic resonance imaging revealed a well-defined pineal region mass causing obstructive hydrocephalus. The patient initially underwent endoscopic third ventriculostomy with biopsy, followed by near-total surgical resection via posterior occipital craniotomy. Histopathological examination confirmed a WHO grade II PPTID, with a low proliferative index [Ki-67 approximately 4%]. Staging investigations, including spinal MRI and cerebrospinal fluid cytology, showed no evidence of dissemination. The patient subsequently received focal adjuvant radiotherapy to the tumor bed [54 Gy in 30 fractions]. Follow-up imaging demonstrated no evidence of disease recurrence. ***Discussion:*** PPTIDs are rare tumors with nonspecific radiological features, making diagnosis challenging prior to histological confirmation. Surgical resection remains the cornerstone of treatment, while adjuvant radiotherapy is associated with improved local control, particularly in cases of incomplete resection. The role of craniospinal irradiation [CSI] remains debated and is generally reserved for patients with cerebrospinal fluid dissemination or high-risk features. In localized, low-grade disease, focal radiotherapy appears sufficient to achieve favorable outcomes. ***Conclusion:*** This case highlights the importance of a multidisciplinary approach in the management of PPTID. Maximal safe resection followed by focal radiotherapy can provide effective disease control in patients without evidence of dissemination. Further studies are needed to establish standardized treatment protocols for this rare entity.

Keywords: Pineal parenchymal tumor of intermediate differentiation, PPTID, Pineal region mass, Adjuvant radiotherapy, Obstructive hydrocephalus, WHO grade II.

INTRODUCTION

Pineal region tumors are rare intracranial neoplasms, accounting for approximately 1% of all central nervous system tumors [1]. They comprise a heterogeneous group of lesions arising from various cellular origins, including germ cells, pineal parenchyma, and supporting glial structures. Among these, pineal parenchymal tumors represent a distinct but relatively uncommon subset, characterized by a broad spectrum of histopathological features and clinical behavior.

According to the World Health Organization [WHO] classification, pineal parenchymal tumors are categorized into pineocytoma [grade I], pineal parenchymal tumor of intermediate differentiation [grade II–III], papillary tumor of the pineal region [grade II–III], and pineoblastoma [grade IV] [2]. Pineal parenchymal tumor of intermediate differentiation [PPTID] occupies an intermediate position between the indolent pineocytoma and the highly aggressive pineoblastoma, both in terms of histology and prognosis.

PPTIDs are most commonly observed in young adults and children and demonstrate variable morphological patterns, reflecting their heterogeneous nature. Radiological evaluation often lacks specificity, making preoperative differentiation from other pineal region tumors challenging. Clinically, patients

frequently present with signs of increased intracranial pressure secondary to obstructive hydrocephalus.

Surgical resection, with or without adjuvant radiotherapy, remains the mainstay of treatment; however, due to the rarity of these tumors, there is no standardized management protocol, and long-term outcomes remain incompletely defined [3,4].

In this context, we present a case of pineal parenchymal tumor of intermediate differentiation and provide a review of the existing literature to further elucidate its clinical, radiological, and pathological characteristics.

CASE PRESENTATION

A 38-year-old previously healthy woman presented to a local hospital with rapidly progressive confusion and headaches, followed by neurological deterioration with the development of aphasia. MRI brain was advised. It showed a lesional process in the pineal region, well-defined with lobulated contours, demonstrating heterogeneous hypointensity on T1-weighted imaging, and mild heterogeneous hyperintensity on T2 and FLAIR sequences, with moderate diffusion hyperintensity. The lesion contains cystic components and shows intense, heterogeneous enhancement after contrast administration measuring 27 × 34 × 21 mm. The lesion exerts significant mass effect, including: Compression of the cerebral aqueduct [aqueduct of Sylvius], Bulging into the third ventricle and Effacement of the quadrigeminal cistern.

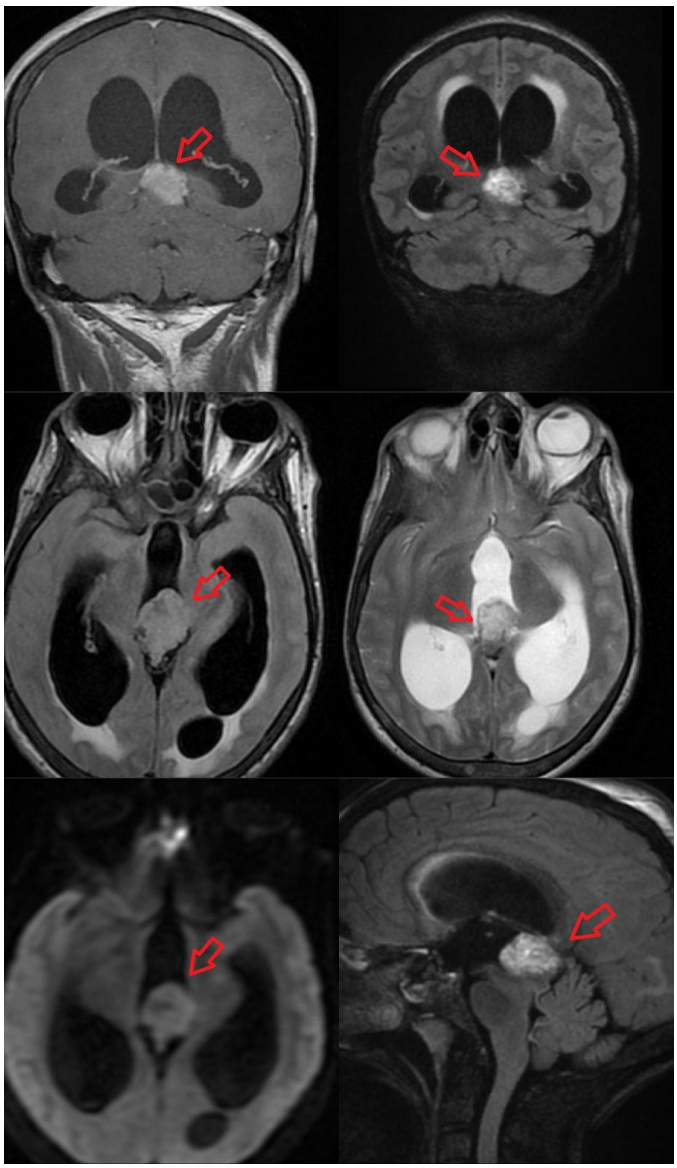

Figure 1: Few selected MR images of the brain at the time of the initial diagnosis demonstrate the well- defined pineal lesion

She underwent a right frontal burr hole and third ventriculostomy with endoscopic biopsy followed by a right posterior occipital craniotomy a month later to excise the pineal mass, which was achieved with at least a near-total resection with post-operative MRI brain showing only minimal patchy enhancement in the tumor bed at pineal region. Postoperative recovery was uneventful, and a sample of the tumor was sent for pathological analysis.

A microscopic examination of the tumor cells showed monomorphic, round/oval nuclei with occasional perinuclear clearing and microcalcification [figure 2]. Tumor cells were immunopositive for CK8/18, S100, and immunonegative for epithelial membrane antigen [EMA]. Additionally, there was focal glial fibrillary acidic protein [GFAP] and synaptophysin immunostaining. Ki-67 immunolabeling was estimated to reach 4%. The tumor was diagnosed as a pineal parenchymal tumor of intermediate differentiation with a WHO grade II.

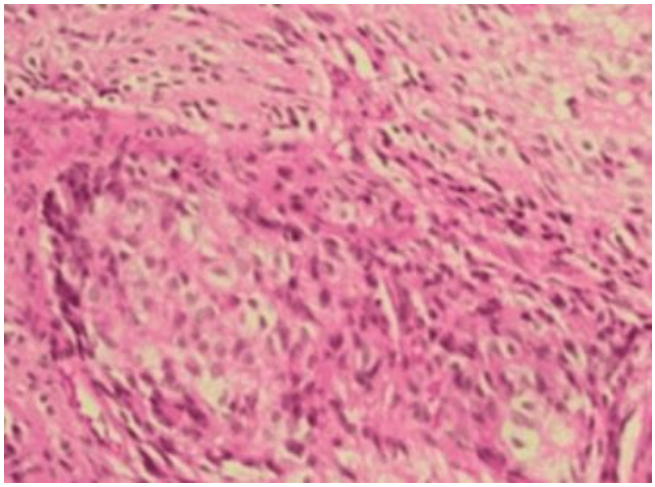

Figure 2: Pathology slide show monomorphic, roud/oval nuclei with occasional perinuclear clearing and microcalcification

MRI pan-spine at initial diagnosis showed no evidence of disease dissemination, which was confirmed by CSF cytology negative for malignant cells 1 month after craniotomy. She subsequently received focused adjuvant RT of 5400 cGy in 30 fractions to the pineal tumor bed [1.8 Gy per fraction]. The radiation was delivered by a linear accelerator with energy of 6 MV. Surveillance MR images of the brain demonstrated no evidence of disease.

DISCUSSION

Pineal region tumors are uncommon, accounting for approximately 1% of all intracranial neoplasms [1]. The majority of these lesions are of germ cell origin, including germinomas, embryonal tumors, and choriocarcinomas [5]. Tumors arising from pinealocytes or their precursor cells constitute the second most frequent subgroup, referred to as pineal parenchymal tumors.

Historically, pineal parenchymal tumors were classified by the World Health Organization [WHO] into two main categories: pineocytomas [grade I] and pineoblastomas [grade IV]. This classification was later refined with the introduction of pineal parenchymal tumor of intermediate differentiation [PPTID] in the 2007 WHO update, representing lesions with features between these two extremes. This revision established a three-tiered histological classification system for pineal parenchymal tumors. PPTIDs are reported to account for a significant proportion, approximately 45%, of these tumors [2].

PPTIDs are most frequently diagnosed in young adults, although they may occur across a wide age range. Clinically, patients commonly present with signs of increased intracranial pressure, such as headache, nausea, and altered mental status, often related to obstructive hydrocephalus secondary to compression of the aqueduct of Sylvius. In the present case, the patient exhibited rapidly progressive confusion and headaches, followed by aphasia, reflecting both raised intracranial pressure and involvement of adjacent neural structures.

Radiologically, PPTIDs lack pathognomonic features, making differentiation from other pineal region tumors challenging. On MRI, these tumors are typically well-defined and may demonstrate iso- to hypointensity on T1-weighted images and variable hyperintensity on T2-weighted sequences, often with heterogeneous contrast enhancement. The presence of cystic components and moderate diffusion restriction, as observed in our case, has been reported but is not specific. Importantly, the significant mass effect with aqueductal compression and resultant hydrocephalus frequently necessitates cerebrospinal fluid diversion prior to definitive surgical management.

Histopathological evaluation remains essential for diagnosis. PPTIDs are characterized by relatively uniform cells with round to oval nuclei, moderate cellularity, and variable mitotic activity. Immunohistochemistry typically demonstrates neuronal differentiation, with positivity for markers such as synaptophysin and variable expression of GFAP, reflecting partial glial differentiation [6]. The Ki-67 proliferation index is a key parameter in grading, with lower values generally associated with WHO grade II lesions. In our case, the low proliferative index [approximately 4%] and morphological features supported the diagnosis of a grade II PPTID.

Surgical resection is considered the cornerstone of treatment, with the goal of achieving maximal safe resection. In recent years, advances in neuro-navigation and microsurgical techniques have improved surgical outcomes in this anatomically complex region. However, due to the deep location and proximity to critical neurovascular structures, complete resection is not always feasible. Adjuvant radiotherapy is commonly recommended, particularly in cases of subtotal resection or higher-grade tumors, to reduce the risk of recurrence. In the present case, near-total resection followed by adjuvant radiotherapy resulted in favorable early outcomes, with no evidence of residual or recurrent disease on follow-up imaging.

PPTID is considered a radiosensitive tumor. Rickert *et al.,* demonstrated that, while PPTIDs share genomic similarities with pineoblastomas, their clinical behavior and prognosis more closely resemble those of pineocytomas. This observation raises an important therapeutic question regarding whether PPTIDs should be managed according to treatment strategies used for pineocytomas or for pineoblastomas [7].

One of the largest metanalyses heretofore conducted incorporating 29 studies and 127 PPTID patients, Mallick *et al.,* found that adjuvant RT significantly improved overall survival [OS] to 252 months compared to 168 months for patients who underwent surgery alone [8]. They further reported that extent of surgery and use of adjuvant chemotherapy did not significantly impact OS.

A retrospective series from the MD Anderson Cancer Center including 17 patients with PPTID provides important insights into the role of adjuvant radiotherapy. Among patients who underwent gross total resection [GTR], those who received postoperative radiotherapy demonstrated improved local control compared to those who did not. Specifically, only one recurrence was observed among patients treated with both GTR and adjuvant radiotherapy, occurring at 16.8 months. In contrast, the single patient who did not receive adjuvant radiotherapy experienced early recurrence at 12.7 months. This patient was subsequently treated with salvage stereotactic radiosurgery [SRS], achieving durable disease control with no evidence of recurrence at long-term follow-up [111 months] [9].

Furthermore, a substantial proportion of patients without cerebrospinal fluid [CSF] dissemination at diagnosis who received radiotherapy were treated with upfront craniospinal irradiation [CSI], and all patients who underwent GTR followed by radiotherapy in this cohort received CSI. These findings highlight the potential benefit of adjuvant radiotherapy in improving disease control; however, they also underscore the variability in radiation strategies, particularly regarding the use of CSI in the absence of CSF dissemination [9]. The use of craniospinal irradiation in patients without cerebrospinal fluid dissemination remains controversial and may represent overtreatment in selected cases.

The association between radiation dose and survival outcome is also a matter of debate. A study by Schild et al [10] of 30 patients showed an link between the radiation dose and survival time in patients with pineal parenchymal tumours. In that study patients who received doses >50 Gy had a significantly improved 3-year survival rate compared to those who received lower doses [94 vs 56 %, respectively].

Despite these therapeutic strategies, the optimal management of PPTID remains controversial due to its rarity and the limited number of reported cases. Long-term prognosis is variable and depends on factors such as extent of resection, histological grade, and proliferative index. Consequently, close radiological surveillance is essential.

This case highlights the diagnostic and therapeutic challenges associated with PPTID and underscores the importance of a multidisciplinary approach combining radiological, surgical, and pathological expertise. Further studies with larger cohorts are needed to better define standardized treatment protocols and prognostic factors for this rare entity.

CONCLUSION

Pineal parenchymal tumor of intermediate differentiation is a rare and heterogeneous entity with variable clinical behavior, posing significant diagnostic and therapeutic challenges. Due to the absence of pathognomonic imaging features, definitive diagnosis relies on histopathological and immunohistochemical evaluation. Management strategies remain controversial, particularly regarding the extent of surgical resection and the optimal use of adjuvant therapies. This case highlights that a multimodal approach combining maximal safe resection and focal radiotherapy can achieve favorable outcomes in patients with localized, low-grade disease without cerebrospinal fluid dissemination. The role of craniospinal irradiation should be carefully considered and reserved for selected high-risk cases. Further studies with larger cohorts are

required to establish standardized treatment guidelines and to better define prognostic factors for this rare tumor.

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