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Case Report

Behind the Pineal Gland: A Case Report on Pineoblastoma in a Teenager

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Abstract

Pineoblastomas are aggressive tumors of the pineal region, primarily affecting children and adolescents, with frequent craniospinal metastasis. MRI plays a key role in diagnosis, surgical planning, and differentiation from other pineal tumors.

This case report describes a 16-year-old male with headaches, nausea, and vomiting, diagnosed with a WHO grade IV pineoblastoma. Imaging revealed typical features, including hypointensity on T1-weighted MRI and restricted diffusion. Treatment involved surgical resection, followed by adjuvant radiotherapy and chemotherapy.

Pineoblastomas typically present with symptoms of intracranial hypertension due to obstructive hydrocephalus. MRI and CT scans help characterize the tumor, showing hypointensity on T1-weighted images, restricted diffusion, and heterogeneous enhancement following gadolinium injection. The tumor often compresses surrounding brain structures, such as the dorsal midbrain, and shows high cellularity. Pineoblastomas also have a tendency for leptomeningeal spread, necessitating full spinal imaging.

The primary treatment is surgical resection, with adjuvant radiotherapy and chemotherapy. Prognosis improves with complete resection and in patients over 5 years old. The use of high-dose induction chemotherapy and autologous hematopoietic stem cell transplantation may improve outcomes, especially in younger patients.

Keywords: *Pineoblastoma; Pediatrics; Brain MRI; Primitive Neuroectodermal Tumor (PNET); Radiotherapy; Chemotherapy; Surgical Resection*

Abbreviations

PNETs: Primary Neuroectodermal Tumors; MRI: Magnetic Resonance Imaging; CT: Computed Tomography; WHO: World Health Organisation; Cho/NAA: Choline/N-Acetyl Aspartate Ration; CSF: Cerebrospinal Fluid; β-HCG: Beta-Human Chorionic Gonadotropin

Introduction

Pineoblastomas represents 25% to 50% of pineal tumors, and are seen more frequently in children and adolescents. These tumors are considered primary neuroectodermal tumors (PNETs) of the pineal region and exhibit aggressive clinical behavior with frequent metastases throughout the craniospinal axis. Standard of care includes maximal surgical resection with adjuvant craniospinal irradiation and systemic chemotherapy, resulting in a median survival time of 16 to 25 months and a 5-year survival rate of 10% [1]. MRI brain imaging remains the gold standard for non-invasive assessment of pineoblastoma, providing high-resolution images that assist clinicians in both diagnostic and therapeutic planning [2].

Given the tumor's aggressive nature, distinguishing pineoblastoma from other pineal masses through imaging alone can be challenging. Thus, MRI findings are often combined with cerebrospinal fluid analysis and biopsy results for a definitive diagnosis [3].

Case Report

We present the case of a 16 years-old male patient with no notable medical history, who was admitted after experiencing four days of headaches, nausea, and intermittent episodes of projectile vomiting. The headaches initially started two months earlier but have recently increased in both frequency and intensity. He denied any limb paresthesia or visual acuity reduction. On physical examination, he displayed no gait disturbances, neurological deficits, or nystagmus.

The initial CT scan revealed a poorly defined hyperdense brain lesion centered on the tectal plate, which required further characterization through an MRI of the brain with and without gadolinium contrast.

MRI of the brain, both with and without Gadolinium injection, revealed a mass in the pineal gland, showing hypointense signals on T1- and T2-weighted images. There was a mass effect on the tectal plate and the dorsal midbrain, remaining distant from the aqueduct and mammillary bodies (Figure 1). The mass showed restricted diffusion and low ADC values (Figure 2), and intense enhancement following gadolinium administration (Figure 3).

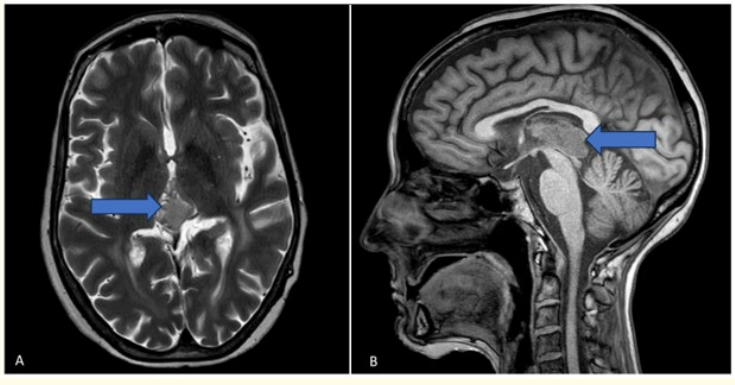

Figure 1: *Pineoblastoma in 16 years old. MRI shows a well-circumscribed uncapsulated tumor, with lobulated contours arising from the pineal region (arrows) (A): Axial T2-weighted image shows that the tumor is intermediate to high signal relative to gray matter. (B) Sagittal T1 image shows that the tumor is isointense to grey matter. In this view, the mass appears compressing the tectal plate and the dorsal midbrain without clear obstruction of the cerebral aqueduct.*

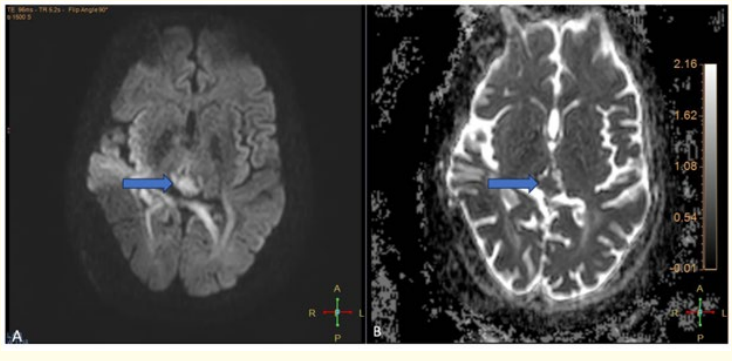

Figure 2: *Pineoblastoma in 16 years old. The tumor shows restricted diffusion as seen on diffusion-weighted imaging (A) and low levels of ADC in the ADC mapping (B). This justify the hypercellularity of the tumor.*

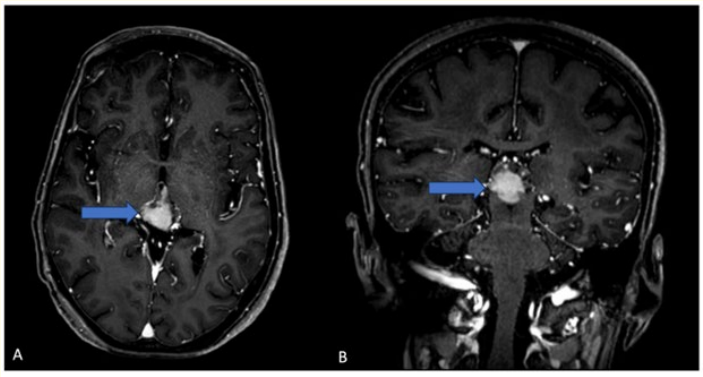

Figure 3: *Pineoblastoma in 16 years old. Axial (A) and coronal (B) T1-weighted gadolinium-enhanced image showed homogeneous and intense enhancement of the tumor (arrows). It is important to note that there is no associated leptomeningeal enhancement.*

An initial lumbar puncture was performed, revealing no significant abnormalities aside from a high opening pressure and borderline protein levels. The patient also underwent tumor markers blood tests and direct CSF measurements (Table 1). Surgical resection was conducted, and histopathological analysis confirmed a papillary pineal tumor, WHO grade IV, also known as pineoblastoma.

CSF*		**Refence ranges**
Appearance	Clear	
Opening Pressure	40 cm H_2O	6-25 cm H_2O
White Blood Cells (WBCs)	2 cells/ µL, primarily lymphocytes	0-5 cells/ µL
Red Blood Cells (RBCs)	absent	0 cells/ µL
Protein	45 mg/dL	15-45 mg/dL
Glucose	50 mg/dL	40-70 mg/dL
Lactate	1,5 mmol/L	1,2-2,1 mmol/L
Chloride	115 mmol/L	110-125 mmol/L
pH	7,33	7,31-7,34
b-HCG*	Undetected	-
AFP*	Undetected	-
Serum		
b-HCG	2 UI/L	< 5 UI/L
AFP	5 ng/mL	< 10 ng/mL

Table 1: *CSF and serum lab results with reference ranges.*

CSF: Cerebrospinal Fluid; b-HCG: Beta-Human Chorionic Gonadotropin; AFP: Alpha-Fetoprotein.

Discussion

Pineoblastoma is an embryonal tumor originating from pineal parenchyma, accounting for 25% to 50% of pineal papillary tumors. It is most commonly observed in children and adolescents, with an average age of diagnosis around 13 years. It represents less than 1% of all primary central nervous system tumors. The World Health Organization classifies it as a grade IV lesion [4].

The clinical presentation typically includes symptoms of intracranial hypertension, resulting from obstructive hydrocephalus of the aqueduct caused by the tumor. Patients may also exhibit Parinaud's syndrome, characterized by paralysis of upward or downward gaze while preserving horizontal movements. This occurs due to compression of the dorsal midbrain, which disrupts the motor pathways responsible for vertical gaze [1]. Physical examination should assess the patient's level of consciousness and check for nystagmus, papilledema, or focal neurological deficits.

The next step involves intracranial imaging, typically a brain MRI with and without contrast. Key imaging findings include a tumor mass greater than 3 cm, with lobulated or irregular contours. It may contain hemorrhagic or necrotic areas, while calcifications are rarely observed. Obstructive hydrocephalus is often present to varying degrees. On a CT scan, the tumor may appear large with blurred edges, exhibiting spontaneous hyperdensity and intense enhancement, which indicates high cellularity. On MRI, pineoblastomas are typically hypointense on T1-weighted images and may contain fat components that appear hyperintense on T1. They show heterogeneous enhancement following gadolinium injection and exhibit restricted diffusion, with the cystic component usually being minimal [5]. Additionally, MRI is useful in depicting local infiltration and leptomeningeal spread [6]. Regarding spectroscopic data, pineoblastoma shows an increased Cho/NAA ratio without a lipid peak [7]. An important distinction among pineal masses is that of a pineal cyst, which is a common benign lesion that appears isointense to cerebrospinal fluid (CSF). Like other pineal tumors, such as germinomas and ependymomas, pineoblastoma has a strong predilection for spinal metastases. Therefore, an injected acquisition of the entire spine should be performed to search for secondary locations. This is followed by blood or direct CSF measurement of specific germ cell tumor markers (β-HCG, gonadotropin, alpha-fetoprotein) to rule out germinomas, which are the most common tumors of the pineal gland [8].

After CSF diversion, the next step is to establish a histological diagnosis, for which several methods can be proposed: stereotactic or endoscopic biopsy. It should be noted that stereotactic biopsy has a higher risk of hemorrhage [9]. Regardless of the method employed, biopsy samples represent only a limited portion of the tumor, potentially missing the diagnosis in 10% of cases [10]. For these reasons, the treatment of choice remains surgical resection.

Adjuvant treatment typically includes a radiotherapy protocol of 5500 cGy directed at the tumor and 3500 cGy for the spine. Chemotherapy protocols are less standardized but often involve 2 or 3 agents, such as vincristine, cisplatin, and cyclophosphamide, among others [9].

Better survival rates were observed in patients over 3 years of age at the time of diagnosis who underwent total surgical resection followed by adjuvant radio-chemotherapy, as well as in those without any secondary locations [11]. A literature review revealed that patients older than 5 years had a better prognosis, with a median survival rate of 57% at 5 years [12]. Therefore, therapeutic management should consider the patient's age. High-dose induction chemotherapy and intrathecal chemotherapy with autologous hematopoietic stem cell transplantation may be proposed for younger children to improve survival outcomes [13].

Conclusion

Pineoblastomas are among the most aggressive tumors of the pineal gland. Due to their malignancy, a thorough and comprehensive assessment is essential. This mass presents itself as a non-encapsulated mass of the pineal region, invading adjacent structures with most often a leptomeningeal dissemination. In our patient, who presented with intracranial hypertension, MRI helped orient the diagnosis by clarifying the invasive and hypervascular nature of the pineal mass found, enabling early intervention and consequently a better prognosis later on.

Conflict of Interest

None.

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