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Case Report

Pineal Gland Epidermoid cyst, a rare Tumor

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ABSTRACT

Background: Intracranial epidermoid cysts, forming 1% of adult and 3-8% of pediatric brain tumors, are among the rarest of intracranial neoplasms. Originating from ectodermal remnants retained during embryonic development, pineal epidermoid cysts are rare benign tumors that often manifest as obstructive hydrocephalus due to midbrain tectum compression.

Objective: To report on the clinical presentation, diagnostic process, and surgical management of a pineal epidermoid cyst in a young adult, emphasizing the role of ventriculoperitoneal shunting and the outcomes of surgical intervention.

Methods: A case study of a 20-year-old male presenting with a 4-month history of occipital headaches and vertigo, without seizures, neurological deficits, or visual disturbances. Diagnostic evaluation included brain imaging that revealed hydrocephalus secondary to a pineal space-occupying lesion. A ventriculoperitoneal shunt was placed for obstructive hydrocephalus treatment prior to the surgical excision of the pineal lesion.

Results: The patient underwent a ventriculoperitoneal shunting procedure, followed by a suboccipital transtentorial craniotomy 2 months later, leading to the complete resection of the pineal space-occupying lesion. The histopathological analysis confirmed the diagnosis of a pineal epidermoid cyst. Post-surgical follow-up indicated significant symptomatic improvement with no recurrence observed.

Conclusion: Pineal epidermoid cysts, while rare, can significantly impact patient well-being through the development of obstructive hydrocephalus. Early intervention with ventriculoperitoneal shunting followed by surgical resection can lead to favorable outcomes. Long-term monitoring is essential to ensure there is no recurrence.

Keywords: Epidermoid Cyst, Hydrocephalus, Intracranial Tumor, Pineal Gland, Surgical Resection, Ventriculoperitoneal Shunt.

INTRODUCTION

Intracranial epidermoid cysts stand as one of the least common varieties of intracranial tumors, encompassing 1% of adult intracranial tumors and between 3% and 8% of pediatric brain tumors (1). Unlike the cerebellopontine angle, which is recognized as the most prevalent site for these cysts, the pineal region represents a notably rare location for their occurrence (2). Interestingly, less than 20% of tumors originating in the pineal region are derived from pineal cells. A pioneering documentation of an intracranial epidermoid cyst within the pineal gland was made by Cushing in 1928, marking a significant milestone in the understanding of these entities. To date, the literature has recorded 85 cases of pineal epidermoid cysts (3, 4). The pineal gland, known for its diverse cell composition, gives rise to a wide spectrum of tumor pathophysiologies in this brain region. This includes germ cell tumors (such as germinoma, yolk sac tumor, choriocarcinoma, embryonal carcinoma), pineocytoma, pineoblastoma, glial cell tumors, meningioma, and both dermoid and epidermoid cysts(5, 6). The preoperative differentiation of pineal epidermoid cysts presents a challenge due to their non-specific symptoms and radiological similarities to other pineal tumors. This report details the case of a 20-year-old male patient who presented with obstructive hydrocephalus as a result of a pineal epidermoid cyst, exploring the clinical presentations, radiological features, and surgical management of this rare condition. The objective of this case report is to enhance the understanding of pineal epidermoid cysts, emphasizing the significance of accurate preoperative identification to facilitate effective treatment strategies(7, 8). This endeavor aims to contribute to the existing body of knowledge, offering insights that may aid in the advancement of diagnostic and therapeutic approaches for such uncommon intracranial tumors.

Journal of Health and Rehabilitation Research 2791-156X JHRR

MATERIAL AND METHODS

Case description/patient history:

A 20-year-old male presented with a four-month history of occipital headaches accompanied by vertigo. These symptoms were not associated with seizures, any focal neurological deficits, or visual disturbances. Upon examination, cranial nerves were found to be intact, and neurological evaluation revealed no focal deficits or cerebellar signs, with a Glasgow Coma Scale (GCS) score of 15/15. Fundoscopic examination did not reveal any papilledema, and the patient had no significant past neurological or ophthalmological history.

Diagnostic workup:

Initial diagnostic efforts included a non-contrast computed tomography (CT) scan of the brain, which identified obstructive hydrocephalus and a hypodense lesion in the pineal region. Further assessment through brain magnetic resonance imaging (MRI) with and without contrast depicted a lobulated, heterogeneous, non-enhancing mass in the pineal region, measuring 2.8x2.6x2.5 cm (CC x AP x TR). The lesion exerted a mass effect, mildly compressing the tectum anteriorly, affecting the cerebellum posteriorly, and impacting the splenium of the corpus callosum superiorly while closely abutting the vein of Galen. These findings were suggestive of an epidermoid cyst, although differentials included pineal region germinoma and teratoma. Serum levels of human chorionic gonadotropin, alkaline phosphatase, and alpha-fetoprotein were assessed to further distinguish between these possibilities(9).

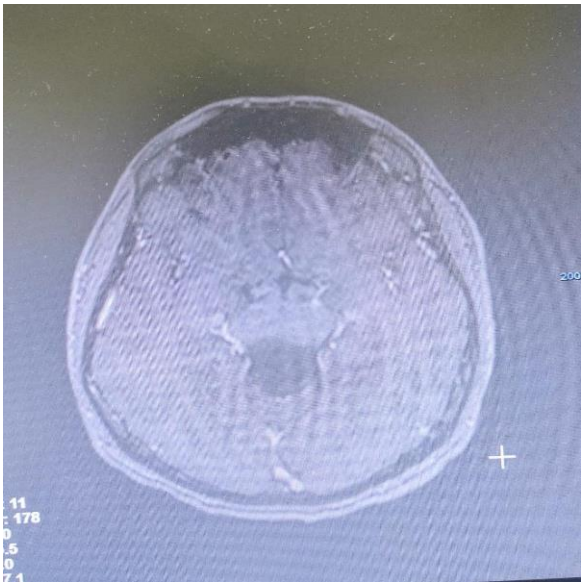
Treatment and outcome:

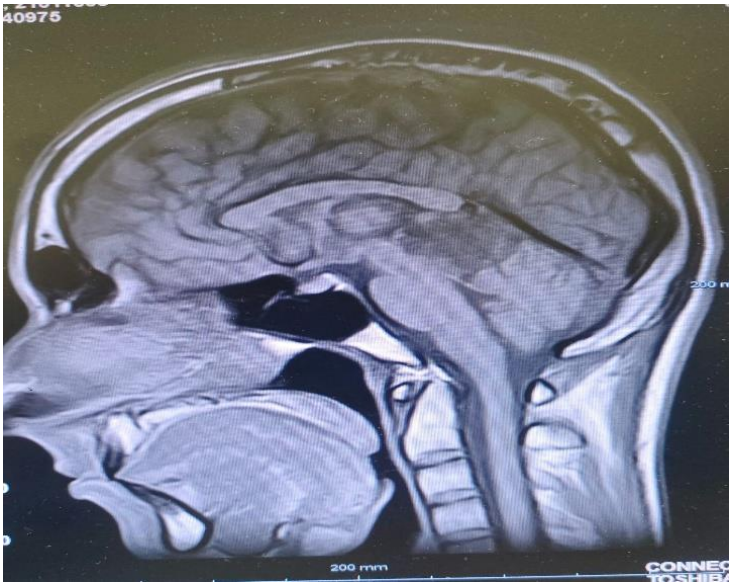
Given the clinical and radiological findings, surgical intervention was recommended. Initially, a ventriculoperitoneal shunt was placed to alleviate the symptoms of obstructive hydrocephalus. Two months later, a suboccipital transtentorial craniotomy was performed under neuronavigation guidance, during which the cyst was carefully dissected and removed from the pineal region, and an external ventricular drain (EVD) was installed. The intraoperative appearance of the lesion, characterized by yellowish, pearly material indicative of keratinous debris and a solid consistency, confirmed the diagnosis of an epidermoid cyst. Histopathological analysis further substantiated this diagnosis(10).

Literature review:

Intracranial epidermoid cysts are among the rarer forms of intracranial tumors, comprising 1% of adult intracranial tumors and 3-8% of pediatric brain tumors (1). The cerebellopontine angle is more commonly affected than the pineal region, where less than 20% of tumors originate from pineal cells (2). Since Cushing's first documentation in 1928, only 85 cases of pineal epidermoid cysts have been reported in the literature (3). The diverse cell types within the pineal gland contribute to the wide variety of tumor pathophysiologies observed in this brain region. Symptoms of pineal cysts can arise from local mass effects or through effects mediated by the third ventricle, including obstruction of the aqueduct of Sylvius, leading to obstructive hydrocephalus, or pressure on the midbrain tectal region, potentially resulting in Parinaud's syndrome. The variable clinical presentations and often vague symptoms of these cysts complicate diagnosis. Contrast CT scans and MRI are pivotal in identifying these lesions, with epidermoid cysts typically exhibiting specific characteristics on imaging modalities. While many pineal cysts remain asymptomatic and clinically inconsequential, consensus dictates that symptomatic individuals, especially those with hydrocephalus, worsening neurological symptoms, or cyst enlargement, should undergo surgical intervention, which can include open surgery, stereotaxy, or neuroendoscopy for tissue biopsy of the pineal lesion.

Journal of Health and Rehabilitation Research JHRR





Ethical approval

Ethical Approval and Patient Consent: Ethical considerations and patient consent were paramount in the preparation of the case report titled "Pineal Gland Epidermoid Cyst: A Case Report." Explicit written consent for publication was obtained from the patient or their legal guardian, encompassing all diagnostic information, clinical history, and images, with thorough anonymization to ensure confidentiality. The medical practices and interventions described were in strict alignment with the highest clinical standards and current medical knowledge, adhering to ethical guidelines including the Declaration of Helsinki. Novel or off-label treatments were pursued only with appropriate approvals, prioritizing patient welfare. This study received ethical approval from Dr. Muhammad Nadeem, Head of the Department of Neurosurgery at Shifa International Hospital, Islamabad, Pakistan, underscoring our commitment to ethical rigor in medical case reporting.

FINDING & DISCUSSION

The pineal gland, a diminutive endocrine gland situated in the brain's midline at the posterior border of the third ventricle, plays a crucial role in regulating sleep-wake cycles through melatonin production. Encased within the posterior cranial fossa, it is juxtaposed with several vital structures, including the third ventricle, habenular nuclei, internal cerebral veins, and the vein of Galen, among others. Dermoid cysts within the pineal gland are an extraordinary occurrence, typically asymptomatic but capable of eliciting symptoms such as headaches, seizures, and vision disturbances when they enlarge and exert pressure on adjacent structures(11).

The rarity of pineal gland dermoid cysts, with only a handful of cases documented in the literature, underscores the uniqueness of the presented case involving a 20-year-old male. This patient's experience of persistent headaches and vertigo without accompanying visual disturbances, seizures, neck rigidity, or focal neurological deficits adds to the diverse spectrum of clinical presentations observed with these cysts. Diagnostic modalities, particularly magnetic resonance imaging (MRI), play an indispensable role in identifying these lesions, offering detailed insights into their size, structure, and the extent of impact on surrounding tissues(1).

Surgical intervention, specifically total surgical resection, emerges as the definitive treatment for pineal gland dermoid cysts. However, the potential for regrowth, even after gross total removal, necessitates a rigorous follow-up regime, extending beyond the duration suggested by prior research to ensure the absence of recurrence. This case's adoption of a suboccipital transtentorial craniotomy, favored for its efficacy in accessing pineal region lesions, highlights the ongoing evolution of surgical strategies to

Journal of Health and Rehabilitation Research 2791-156X JHRR

optimize patient outcomes. The subsequent placement of an external ventricular drain (EVD) underscores the surgical procedure's dual objectives: to alleviate raised intracranial pressure and to monitor cerebrospinal fluid (CSF) characteristics post-operatively.

This discussion accentuates the intricate balance between advancing medical understanding and addressing the practical challenges encountered in managing rare neurological conditions. The strength of this case report lies in its detailed account of the diagnostic and therapeutic journey, contributing valuable insights into a scarcely reported condition. Nevertheless, the limitations inherent in dealing with such rare entities include the constrained ability to draw broad generalizations from single case studies and the ongoing need for longitudinal data to ascertain the long-term efficacy of treatment modalities. Future research endeavors should aim to broaden the understanding of pineal gland dermoid cysts, exploring innovative diagnostic techniques and surgical approaches to enhance patient care in these complex cases.

CONCLUSION

Dermoid cysts of the pineal gland, while uncommon, have the potential to induce substantial symptoms upon enlargement. The preferred method of treatment, surgical resection, has been shown to markedly alleviate these symptoms. It is imperative, however, to engage in vigilant long-term monitoring post-surgery to prevent any recurrence of the cyst.

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