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Cholesteatoma of the Pineal Region of the Brain

[1]Aliyev Mansur Abdukhalikovich, [2]Najimova Margiyona Kakhramonovna, [3]Abdukarimova Zalina Adkhamovna, [4]Shamsiyeva Raykhona Ozodovna

[1]Head of the Department of Neurosurgery, Associate Professor, Phd, [2.3.4]Students of Samarkand State Medical University, Faculty of Pediatrics, Group 229

Abstract: Pineal region cholesteatoma (intracranial epidermoid cyst) is a rare benign congenital lesion characterized by accumulation of keratinized epithelium. Due to its slow growth, the tumor may remain asymptomatic for a long period and becomes clinically evident after compression of surrounding brain structures and ventricular system.

This article presents a clinical case diagnosed using MRI and treated surgically. Neuroimaging revealed a large cystic formation in the pineal region. Microsurgical removal of the lesion was performed successfully, and the postoperative period was without complications.

Early radiological diagnosis and timely surgical intervention are essential for favorable outcomes. Long-term follow-up is recommended because recurrence may occur.

Keywords: cholesteatoma, pineal region, brain, diagnosis, surgical treatment, neurosurgery, complications, recurrences.

Introduction

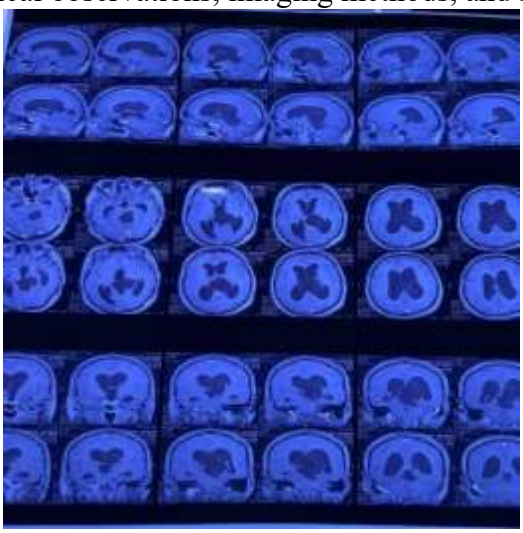
Intracranial epidermoid cysts, historically referred to as cholesteatomas, are rare benign congenital lesions that originate from ectodermal inclusions during the early stages of neural tube closure. These lesions account for approximately 0.2–1% of all primary intracranial tumors and are most commonly located at the cerebellopontine angle, parasellar region, and skull base. Localization within the pineal region is exceptionally uncommon and presents unique diagnostic and surgical challenges due to the deep anatomical location and proximity to vital neurovascular structures, including the midbrain, thalamus, deep venous system, and ventricular pathways. Histologically, epidermoid cysts consist of stratified squamous epithelium and contain keratin debris and cholesterol crystals, which give the lesion its characteristic pearly appearance during surgical removal. Because of their extremely slow growth and non-invasive biological behavior, these tumors may remain clinically silent for many years and are frequently discovered only after they reach a considerable size. Clinical manifestations usually result from mass effect and obstruction of cerebrospinal fluid circulation, leading to headaches, visual disturbances, Parinaud syndrome, hydrocephalus, or cognitive changes.

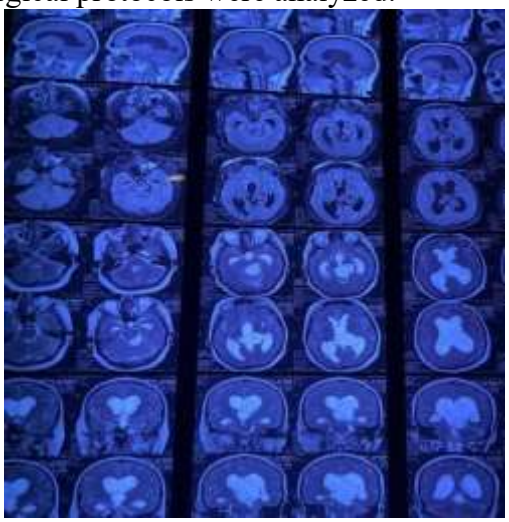
Microsurgical excision remains the treatment of choice and aims at maximal safe removal while preserving critical venous and neural anatomy. However, total removal of the capsule is often difficult, and residual fragments may cause late recurrence even years after surgery, making longterm follow-up mandatory. The rarity of pineal epidermoid cysts and the complexity of their

surgical management highlight the importance of presenting clinical observations and analyzing diagnostic and therapeutic approaches to improve treatment outcomes and neurosurgical practice.

Materials and Methods

The article analyzes clinical cases published in world literature, as well as diagnostic data from MRI and CT imaging. Statistical data on disease prevalence and treatment effectiveness were reviewed. Clinical observations, imaging methods, and surgical protocols were analyzed.





Patient Information

Patient Name: Oripova Gulbahor

Year of Birth: 1975

Date: 22.01.2025

MRI revealed a cystic lesion in the pineal region compressing the brain ventricles, measuring 72 × 61 × 55 mm, with relatively clear contours. The lesion showed heterogeneous intensity on T2weighted and FLAIR images.

Results and Discussion

Magnetic resonance imaging revealed a large cystic mass located in the pineal region measuring approximately 72 × 61 × 55 mm, causing compression of the ventricular system and displacement of surrounding brain structures. The lesion demonstrated heterogeneous signal intensity on T2weighted and FLAIR sequences, which is typical for intracranial epidermoid cysts due to the presence of keratin and cholesterol components. Ventricular dilatation indicated developing obstructive hydrocephalus, explaining the progressive intracranial pressure effects. The relatively well-defined margins and absence of solid contrast-enhancing components allowed differentiation from malignant pineal tumors such as germinoma or pineoblastoma. Based on neuroimaging findings, a space-occupying benign lesion was suspected and surgical treatment was recommended.

A left parietotemporal craniotomy was performed with microsurgical removal of the intraventricular mass involving the lateral ventricles, third ventricle, and pineal region. Intraoperatively, the lesion had a pearly white appearance with soft keratinous content, consistent with an epidermoid cyst. Careful dissection was required because the capsule showed adhesions to surrounding neural structures and ventricular walls. Maximal safe removal was achieved while preserving adjacent brain tissue and deep venous structures. The postoperative period was uneventful, and no immediate neurological deficits were observed, confirming the effectiveness of early surgical intervention in preventing irreversible neurological damage.

From a clinical perspective, pineal region epidermoid cysts often remain undiagnosed until they reach a large size because symptoms appear gradually and are primarily related to compression rather than invasion. In many reported cases, hydrocephalus and visual disturbances are the first manifestations. MRI plays a crucial role in distinguishing epidermoid cysts from arachnoid cysts, which typically follow cerebrospinal fluid signal intensity and do not show heterogeneous internal structure. The presence of irregular internal signal characteristics and typical intraoperative appearance strongly support the diagnosis. Literature data indicate that complete capsule removal may not always be feasible due to adherence to vital neurovascular structures, and aggressive attempts at total excision increase the risk of neurological complications. Therefore, modern neurosurgical strategy emphasizes maximal safe resection rather than radical removal.

The favorable postoperative outcome in this case is consistent with published observations showing that epidermoid cysts have good prognosis after microsurgical decompression. Nevertheless, recurrence remains possible because residual epithelial fragments can continue keratin production. For this reason, long-term clinical and radiological monitoring is essential even in asymptomatic patients. Early diagnosis combined with careful microsurgical technique significantly reduces morbidity and improves quality of life, highlighting the importance of timely neuroimaging and individualized surgical planning in patients with deep intracranial benign lesions.

Conclusion

Cholesteatomas can destroy bone tissue and progressively damage adjacent structures. Gradual intracranial involvement may not cause acute symptoms, making diagnosis challenging. Detailed clinical and radiological examinations are essential. Lifelong follow-up is required due to possible delayed recurrences.

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