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Case Report

Beneath the surface: unravelling a frontal epidermoid cyst–a case study

Arwa M. Jamali[1]*, Rakeshkumar Luhana[2]

[1]GMERS Medical College, The Maharaja Sayajirao University of Baroda, Vadodara, Gujarat, India
[2]Department of Neurosurgery, Venus Superspecialty Hospital, Vadodara, Gujarat, India

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***Correspondence:**
Dr. Arwa M. Jamali,
E-mail: jamaliarwa123@gmail.com

ABSTRACT

Epidermoid cysts are rare, benign congenital lesions that account for a small percentage (0.3–1.8%) of primary intracranial tumors. These cysts arise from ectodermal cells that become trapped during the early closure of the neural tube in embryonic development. But their occurrence in the frontal lobe is extremely rare, with about 20 cases reported in literature. A 42-year-old male with a history of hypertension presented with generalized tonic-clonic seizures. magnetic resonance imaging (MRI) of the brain revealed a right frontal lobe lesion with mass effect, characterized as a non-enhancing intra-axial lesion. This imaging led to a preoperative diagnosis of low-grade glioma. The patient underwent a right frontal craniotomy with total microsurgical excision of the lesion and duroplasty under general anesthesia. Histopathological analysis of the lesion confirmed it to be an epidermoid cyst. Intracerebral epidermoid cysts, especially those located in the frontal lobe, are exceedingly rare and pose diagnostic challenges due to their similarity to more common brain lesions such as gliomas. Advanced imaging techniques, particularly diffusion-weighted MRI, are crucial in differentiating these cysts from other intracerebral tumors. Complete surgical resection is the definitive treatment, effectively preventing recurrence. The rarity of frontal lobe epidermoid cysts underscores the importance of thorough diagnostic and surgical strategies in managing this unique case.

Keywords: Epidermoid cyst, Frontal lobe tumor, Intracerebral lesion

INTRODUCTION

Epidermoid cysts are benign congenital inclusion accounting for about 0.3–1.8% of primary intracranial tumors.[1] The formation of these infrequent intracerebral abnormalities were from cells entrapment from the mesoectodermal origin of the neural crest within the primitive cerebral hemisphere.[2] The tumor could potentially manifest at various locations. Approximately 90% of intracranial epidermoid cysts are located in the intradural compartment, although they can also occur extradurally in the intradiploic space of the frontal, parietal, temporal, and occipital bones. Epidermoid cysts are most commonly found in the cerebellopontine angle (approximately 40% of all cases), the parasellar region (30%), and the fourth ventricle (5–18%) and less commonly in the middle cranial fossa, diploe or spinal canal.[3] Only few cases of parenchymal epidermoid cysts have been reported. Frontal lobe epidermoid cyst is extremely unusual.[4] The presence of such cysts in the frontal region might lead to specific neurological symptoms depending on their size and proximity to vital brain structures. Surgical intervention is often considered for the removal of these cysts if they cause symptoms or if there is concern about their potential impact on brain function. Consequently, comprehending the presentation, diagnosis, and treatment is essential for prompt and efficient care. Due to the scarce available literature, this case report aims to provide valuable perspectives by exploring these dimensions. This endeavor seeks to enhance the existing understanding of this uncommon occurrence, ultimately fostering better insights into its characteristics and better practices for its management.

CASE REPORT

A 42-year-old male patient was admitted to the hospital with episode of generalized tonic-clonic seizures. Patient is a known case of hypertension on regular medication. No previous history of similar complain and denied any history of head trauma or neurological disorders. No history of ischemic heart disease (IHD) or cerebrovascular attack was mentioned. Family history for similar symptoms was negative. On examination BP=100/70 mmHg, pulse=93/min, SpO_2=99%, Temperature=97. Neurologic examination revealed no abnormality. MRI scan brain showed right frontal lobe with mass effect, intra-axial non enhancing lesion (Figure 1). With these imaging findings, a preoperative diagnosis of low-grade glioma was made.

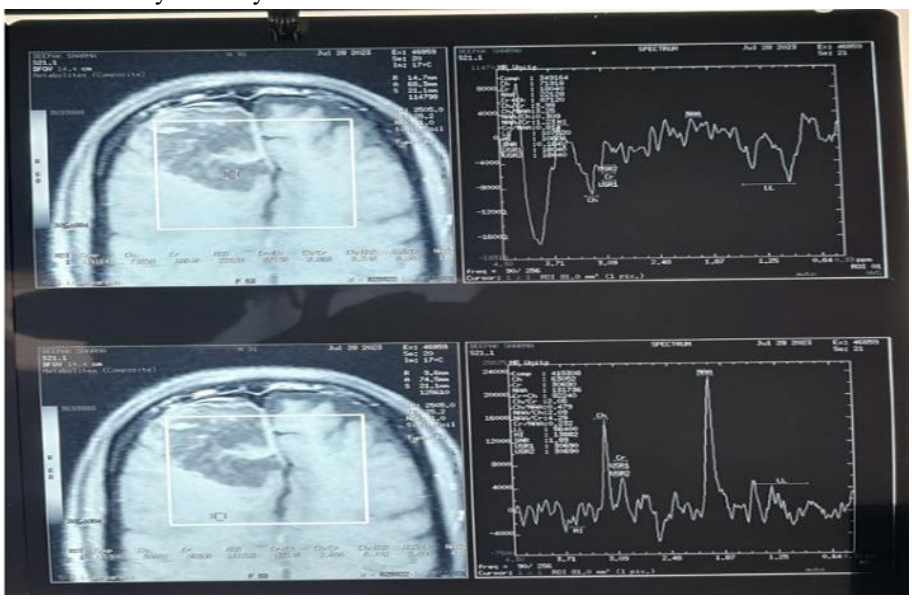

Figure 1: MRI image showing lesion in right frontal lobe.

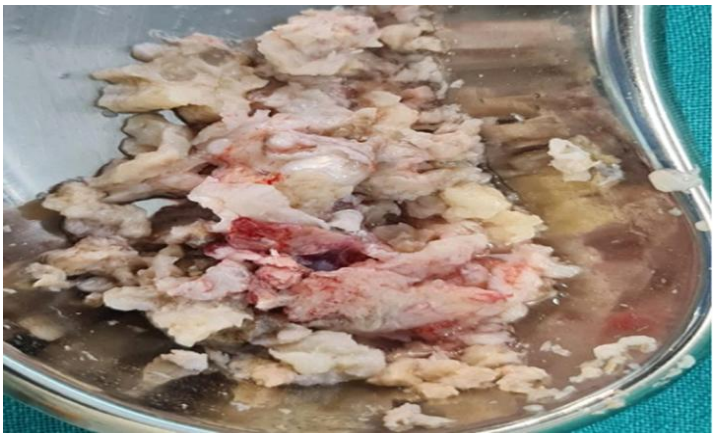

Figure 2: Post-excision specimen containing lesion tissue.

Right frontal craniotomy with total microsurgical excision of right frontal space occupying lesion and duroplasty was done under general anaesthesia (Figure 2).

Intra-op course was uneventful and post-op patient was kept in intensive care unit (ICU) for observation. Patients was stable post-op, and the symptoms did not reoccur. Following surgery, histopathology of the lesion revealed features of epidermoid cyst, namely, keratinizing squamous epithelium arranged in laminated layers with a thin layer of fibrous connective tissue. Patient was given cefuroxime axetil (500 mg), levetiracetam (500 mg), and metoprolol (50 mg). On follow up, the patient reported no recurrence of symptoms and remained clinically stable. A repeat MRI showed no evidence of residual or new lesion. These findings suggest resolution of the previous pathology.

DISCUSSION

Epidermoid tumor is a rare, lesion with benign nature and course. Intracerebral location has been infrequently documented and accounts for about 1.8% of all primary intracranial tumors.[1] These tumors originate from subset

of cells that stem from mesoectodermal lines responsible for skin development that might migrate and become confined within the early cerebral hemisphere during the closure of the neural tube in the 4th and 5th weeks of embryogenesis. These confined cells could undergo development and specialization, giving rise to an epidermoid tumor.[2] Epidermoid tumors exhibit a pearly white appearance, resulting from the buildup of keratin and cholesterol crystals. These substances are derived from the disintegrating surface epithelial cells. The cyst's external capsule comprises the epithelial layer, while the interior contains cystic fluid.[1] Categorized according to their location, they can be classified into four groups: retrocerebellar within the cerebellopontine angle, parasellar within the sylvian fissure, suprasellar near the chiasmatic region, and basilar within the posterior fossa, ranging from the most prevalent site to the least common.[2,5] Location in the frontal lobe is extremely rare , with about 20 cases reported in literature.[6]

As they tend to grow primarily along the cisternal areas of the brain, their clinical presentation and subsequent diagnosis can be significantly delayed. They often remain asymptomatic for a long period and present with symptoms associated with their mass effect once they have reached a considerable size. Common symptoms that arise include persistent headaches, deficits in cranial nerves, manifestations related to the cerebellum, seizures, and an increase in intracranial pressure. While recurrent aseptic meningitis may also occur, it is a relatively infrequent phenomenon. The development of hydrocephalus is quite rare due to the tumor's gradual growth pattern.[7,8] It is a slow-growing tumors with a higher incidence in the fourth decade of life with a male predominance as was also seen in our case.[8]

Key differential diagnoses for epidermoid cysts encompass dermoid cysts, glioma, arachnoid cysts, hamartomatous, lipomas, neurocysticercosis, neuroenteric cysts, and various cystic neoplasms.[9] Nevertheless, the utilization of MRI scans and incorporating techniques such as fluid attenuated inversion recovery (FLAIR), constructive interference in steady state (CISS) sequences, and diffusion-weighted imaging (DWI) could assist in distinguishing intracerebral epidermoid tumors from other types of cystic intracerebral tumors. On FLAIR sequences, intracerebral epidermoid tumors exhibit moderate hyperintensity, and on CISS sequences, they appear hypointense. This distinction aids in enhancing the precision of preoperative diagnoses. However, it's important to note that the signal intensity observed in FLAIR and CISS sequences for epidermoid tumors can resemble those of astrocytomas and arachnoid cysts. To further refine the preoperative radiological assessment of epidermoid tumors, diffusion-weighted imaging is valuable, revealing a distinct and pronounced hyperintense signal.[10,11] Surgical removal remains the primary approach for treatment. Meticulous microscopic dissection of the entire capsule is crucial to prevent recurrence. In cases of partial removal, recurrence can occur over a prolonged period due to the tumor's slow growth rate. However, for specific instances involving significant attachment to neighboring brain tissue or critical vessels, particularly in sensitive brain regions, it is recommended to retain small segments of the tumor capsule. This approach aims to reduce potential postoperative complications.[12] Maintaining the integrity of the capsule during the removal procedure is essential to avoid leakage of cyst contents, as this could lead to the development of chemical meningitis. If, however, the capsule ruptures, promptly administering corticosteroids could be beneficial. Given the tumor's benign characteristics, the patient prognosis is excellent unless specific complications arise during and after the surgical procedure. Postoperative, diligent follow-up is vital for prompt detection of potential recurrence. MRI stands as the most effective diagnostic test for this purpose.[9]

The single-patient focus of this case report and the relatively short follow-up period limit the generalizability of findings and long-term outcome assessment. The report lacks genetic insights and may not encompass diverse treatment approaches. Future studies should conduct extended follow-up on larger cohorts, compare treatment modalities, and incorporate advanced imaging techniques.

CONCLUSION

Intracerebral epidermoid tumors particularly in the frontal lobe are very rare lesions. Its accurate preoperative diagnosis may pose challenges because of its close resemblance to more prevalent intraparenchymal cystic tumors such as astrocytomas or gliomas. Diffusion-weighted MRI may help in the final preoperative diagnosis. Currently, discussions revolve around the most effective approach to treating epidermoid cysts. Complete removal of the lesion through gross total resection is the definitive solution, which effectively prevents recurrence and aseptic meningitis. However, in situations where the tumor capsule strongly attaches to nearby brain tissue, a more cautious approach could be advisable, especially in vital brain regions.

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