


Atypical Obstructive Hydrocephalus Induced by Content Leakage from a Lateral Ventricle Epidermoid Cyst: A Case Report

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Abstract

This report describes an unusual case of biventricular involvement caused by an epidermoid cyst in a 60-year-old man who presented with symptoms such as headache, dizziness, nausea, vomiting and gait ataxia. Investigations for infectious diseases and metastatic neoplasms were negative. Magnetic resonance imaging (MRI) of the brain revealed an expansive intracranial lesion with a cystic appearance in the left lateral ventricle, as well as similar material in the fourth ventricle associated with hydrocephalus. Surgical removal of the fourth ventricle mass restored normal cerebrospinal fluid (CSF) drainage, resulting in significant resolution of the hydrocephalus and improvement of symptoms.

Keywords

Magnetic Resonance Imaging, Neoplasm, Epidermoid Cyst

1. Introduction

Epidermoid cysts are congenital, non-neoplastic brain lesions, accounting for less than 2% of all intracranial tumors [1] [2]. Other cystic lesions in this category include dermoid, arachnoid, and colloid cysts. These cysts develop during neural tube closure when ectodermal cells fail to migrate properly, resulting in remnants of epidermal tissue that grow similarly to other body tissues. Due to their slow

growth, epidermoid cysts often remain asymptomatic until the fifth decade of life [1] [3].

When symptoms manifest, they are typically caused by mass effect, or, in severe cases, by aseptic meningitis following cyst rupture, exposing keratin and cholesterol byproducts. These substances are radiologically seen as an avascular, well-defined capsule containing a pearly white mass, which has led to alternative nomenclature such as “cholesteatoma” or “pearl tumor.”

Epidermoid cysts are typically located off the midline, with the majority occurring in the cerebellopontine angle (CPA) [4] [5]. However, they can also be found in other regions, including parasellar areas. Ventricular involvement is rare, with few documented cases. The simultaneous involvement of both the fourth and lateral ventricles is exceptionally rare and has not been previously described. This case report aims to present the clinical presentation, radiological findings, and management of this unusual case, contributing to the limited literature on epidermoid cysts in the ventricular system [6] [7].

2. Case Description

This manuscript was configured within the ethical aspect of anonymity and without opposition from the patient in reporting his case.

A 60-year-old man from a rural area was transferred to the state’s referral hospital for neurological changes about a month ago, including headache, dizziness, nausea, vomiting and gait ataxia. On admission, the patient had difficulty getting around on his own, difficulty standing, slurred speech and some degree of mental confusion. The patient had previously denied similar symptoms.

No abnormalities in cranial nerve function, strength or reflexes were observed. Serological and cerebrospinal fluid (CSF) tests for infectious diseases were negative. Computed tomography (CT) scans of the chest and abdomen did not reveal any possible sites of primary neoplasia. Initially, a CT scan of the cranial region showed hydrocephalus of the supra- and infratentorial ventricular system associated with an expansive lesion with a cystic appearance, with defined limits, in the temporoparietal region, with an estimated volume of 111.7 cm³, in close contact with the posterior horn of the lateral ventricle/choroid plexus and with foci of marginal calcifications.

For better etiological characterization, cranial MRI was indicated, which identified a cystic lesion in the left temporoparietal region, measuring 7.7 × 5.8 × 5.3 cm, hypointense on T1-weighted images and hyperintense on T2-weighted images. The lesion was in close contact with the left lateral ventricle, showing minimal contrast enhancement in the medial and anterior walls, with no significant edema or mass effect. (**Figure 1**) Amorphous material with similar radiological characteristics to the cyst contents was noted in the fourth ventricle, resulting in stenosis of the Luschka and Magendie foramina, causing hydrocephalus and dilation of both supra- and infratentorial ventricular systems. Based on these findings, the primary hypothesis was that the obstruction of the fourth ventricle was due to leakage of cystic contents from the lateral ventricle, with hydrocephalus being the

primary cause of the symptoms rather than the mass effect of the tumor. (**Figure 2**)

The neurosurgical approach was scheduled to be carried out in two stages, initially draining the lateral ventricles to reduce intracranial pressure and relieve the patient's symptoms, as well as removing the mass from the fourth ventricle, as it was believed to be the main contributor to the hydrocephalus. The material removed during surgery was sent for histopathological analysis, and an excision of the expansive mass would be scheduled at a later date.

This approach was chosen to avoid exacerbating symptoms by manipulating both lesions, which could have induced aseptic meningitis. Endoscopic third ventriculostomy was not feasible due to anterior and inferior displacement of the cinical tubercle. Histopathological examination of the extracted tissue confirmed the diagnosis of an epidermoid cyst. After four days, the external ventricular drain was removed, and there were no signs of hydrocephalus on the postoperative cranial CT scan, with subsequent improvement in symptoms.

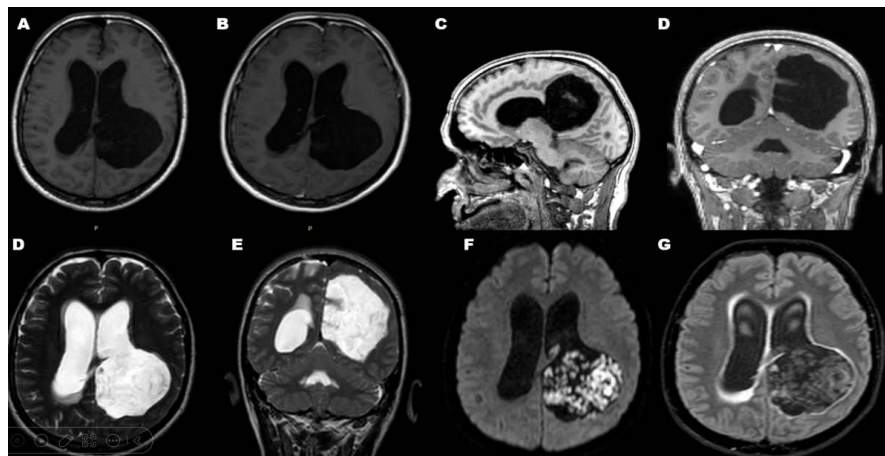


Figure 1. MRI revealing a large intra-axial cystic lesion in contiguity with the left lateral ventricle, compatible with an epidermoid cyst. T1-weighted axial view (A). T1-weighted post-contrast axial view (B). T1-weighted sagittal view (C). T1-weighted coronal view post-contrast (D). T2-weighted axial and coronal views (E and F). Diffusion-weighted axial view (G). Axial flair image (H).



Figure 2. MRI with contrast revealing deposition of neoplastic debris in the floor of the fourth ventricle, determining obstruction of the foramina of Luschka and Mangendie and causing hydrocephalus. Axial flair image (A). T2-weighted axial view (B). Diffusion-weighted axial view (C).

Unfortunately, despite being informed about the risks of abandoning treatment, the patient, in this case, requested to be discharged from the hospital for his own reasons before the second-stage surgery to remove the cystic formation was carried out. The patient then signed a disclaimer stating that he had been informed and was aware of the risks of abandoning the procedure.

3. Discussion

This report presents a 60-year-old male without prior comorbidities who developed symptoms including headache, dizziness, nausea, vomiting, and gait ataxia over one month. MRI revealed an expansile intracranial lesion, and investigations of infectious disease and metastatic neoplasms were negative [6]. On CT, epidermoid cysts typically appear as hypoattenuating lesions without enhancement, although minimal rim enhancement has been observed. On MRI, these cysts are typically hypointense on T1-weighted images and hyperintense on T2-weighted images, a common feature, although variations may occur depending on the cyst composition. High-protein epidermoid cysts, or “white epidermoid cysts,” can appear hyperintense on T1 due to elevated albumin levels and hypointense on T2 due to increased viscosity [2] [9] [10].

Epidermoid cysts account for around 1% of all intracranial tumors. They are predominantly congenital (arising from ectodermal inclusion during neural tube closure), but can be acquired through post-surgical implantation or post-trauma. Patients usually discover them between the ages of 20 and 40, as they are slow-growing and take decades to develop symptoms [2] [3] [7].

They have a thin capsule of stratified squamous epithelium. Macroscopically, they appear white and pearly and can be smooth, lobulated or nodular. Internal desquamated epithelial keratin and cholesterol crystals form a malleable, putty-like material. This explains the way the lesions insinuate themselves and surround nerves and arteries with little displacement, as well as their distinctive MRI characteristics. Their most common location is intradural (90%), pontocerebellar angle (40% - 50%), suprasellar cistern (10% - 15%), fourth ventricle (17%) and extradural (10%) [3] [6] [8].

Regarding differential diagnosis, arachnoid and dermoid cysts are the primary considerations. Arachnoid cysts are generally hypo- or isointense on DWI and FLAIR compared to CSF, while dermoid cysts, which contain lipidic material and occasionally hair or glands, are more heterogeneous and typically midline [8] [10]. The patient’s symptoms align with the literature, where lesions in the fourth ventricle often present with similar clinical manifestations [3] [6] [8].

The slow evolution of the expansive lesion, in this case, means that treatment can be carried out conservatively or surgically in symptomatic patients. In many cases, the lesion is firmly attached to neural tissue, including cranial nerves and vessels, making complete resection difficult. The prognosis in cases of subtotal resection is correlated with the patient’s age at the time of diagnosis, probably due to the greater probability of adhesion to vital structures [8] [9]. Recurrence is not

uncommon, although growth is generally slow, and many years can pass without new symptoms [10].

Inaccessible lesions can be aspirated, but extravasation of the contents can increase the risk of complications such as aseptic meningitis, a known risk following the rupture of this cyst, whose treatment usually involves corticosteroids to control the inflammatory response. Symptoms related to mechanical obstruction due to leakage of the cyst's contents, however, have not been described previously, highlighting the rarity of this case [1] [3] [7].

Cranial nerve dysfunction tends to be associated with the duration of symptoms rather than the age at diagnosis, which is consistent with this patient's brief clinical presentation [9]. Although total resection is preferable to avoid recurrence or granulomatous response from residual cyst contents in the CSF, it is not always feasible due to adhesion to delicate structures [5] [10]. However, partial resection is generally associated with favorable outcomes due to the slow growth of the tumor, and long-term management of CSF drainage is recommended. Regular follow-up with MRI is essential for monitoring [8] [9].

Unfortunately, this study encountered significant limitations after this patient lost his hospital follow-up and was discharged, even against medical advice, without having finished his definitive treatment. This prevented us from following up on this unique case in more detail, with more information about his clinical evolution, skull imaging tests, recurrence of the lesion and how he would recover after neurosurgery to remove the lesion.

4. Conclusion

Although the intracranial prevalence of the epidermoid cyst and its differential diagnoses are not so uncommon, the uniqueness of this case is reflected in the unusual location of the epidermoid cyst, the clinical symptoms and the presence of associated supra- and infratentorial hydrocephalus, determined by the mechanism of obstruction caused by the tumor content impacting on the floor of the fourth ventricle, probably due to a rupture and extravasation of the contents of the primary lesion. Due to the rarity of this condition and the limited literature on the subject, this case report could serve as a source of citation and reference for future studies. The patient's successful recovery without symptom recurrence, even without removal of the intracranial lesion, may support more conservative treatment options in cases where total resection is not possible.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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