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Case Report

Intracranial Dermoid Cyst in the Posterior Fossa: A Case Report

Li Li, 1 Seidu A. Richard^{1, 2}, Zhigang Lan^{1*}, Yuekang Zhang¹

¹Department of Neurosurgery, West China Hospital, Sichuan University; 37 Guo Xue Xiang Road, Chengdu, Sichuan, 610041. P. R. China

²Department of Medicine, Princefield University, P. O. Box MA 128, Ho-Volta Region, Ghana, West Africa

Corresponding authors: Zhigang Lan, E-mail: 158075478@qq.com

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ABSTRACT

Introduction: Intracranial dermoid cysts are benign, ectopic squamous epithelial cysts often compose of dermal structures like hair follicles, sweat glands as well as sebaceous glands. This lesions constitutes about 0.5% of all intracranial neoplasms. Thus, the occurrence of a dermoid cyst in the posterior fossa is very rare. We report a rare case of intracranial dermoid cyst in posterior cranial fossa. Case Presentation: Our first case was a 32 years old woman who presented with headaches and dizziness with no nausea, vomiting or fever. CT scan revealed a mass at occipital cistern consistent with a cystic lesion. MRI also revealed an irregular lesion in the posterior part of the medulla oblongata with enhanced edges signifying calcifications. We attained total resection of the tumor in a piece meal approach via surgery. Histopathology confirmed dermoid cyst. Two years follow-up revealed no recurrence of the lesion and no neurological deficits. Conclusion: We advocate that, the goal in surgical decision-making should be safe and total resection while monitoring the cranial nerves with electromyographic and auditory brainstem responses.

INTRODUCTION

Intracranial dermoid cysts are benign, ectopic squamous epithelial cysts often compose of dermal structures like hair follicles, sweat glands as well as sebaceous glands (1, 2). These lesions are congenital malformations that account for about 0.5% of all intracranial neoplasms (3). These lesions often originate from ectopic epithelium sequestered inside the neural tube during embryogenesis (3-8). They have strong predilections for the Sylvian fissure, sellar region as well as cerebellar vermis although isolated cases have been found in the anterior, middle, and posterior cranial fossa at comparable rates (2, 3, 9, 10). Currently only few cases of dermoid cysts have been reported at posterior fossa (2, 11). Furthermore, ruptured posterior fossa dermoid cyst presenting with hydrocephalus as complication have been observed (2, 11).

The cardinal symptomatology of intracranial dermoid cysts are headaches and visual disturbances (3, 4, 9, 10). Radiologically, intracranial dermoid cysts characteristically present as non-enhancing low-density masses on Computer tomographic (CT) scan as well as hyperintense on T1-weighted magnetic resonance imaging (MRI) sequences

with inconsistent signals on T2- weighted sequences (5, 12). The gold-standard treatment modality is surgery (3-8, 12). Surgical approach is often determined by the location of the lesion (4, 12). Intraoperatively, careful dissection of the cyst capsule and neighboring neurovascular structure is often advocated (12). The occurrence of a dermoid cyst in the posterior fossa is very rare. Thus, we report a rare case of intracranial dermoid cyst in posterior cranial fossae.

CASE PRESENTATION

A 32 years old woman presented with a month history of Headaches and dizziness with no nausea, vomiting or fever. She had myomectomy 2 years prior to this presentation. Cranial nerve examinations were unremarkable. General physical examination did not yield much. Routine laboratory investigations were grossly at normal rangers. Routine Chest-X ray and electrocardiogram (ECG) were essentially normal. CT scan revealed a mass measuring about 3.1 x 2.5mm at occipital cistern consistent with a cystic lesion (Figure 1a). All other structures were essential normal. Computer tomographic angiograph (CTA) showed dilated blood

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vessel around the lesion in the occipital cistern. Also, MRI revealed an irregular lesion in the posterior part of the medulla oblongata measuring about 2.4 x 2.3 x 2.2 cm with enhanced encapsulated edges (Figure 1 b-d). The MRI findings were consistent with the intracranial dermoid cyst. Thus, we decided to operate.

After general anesthesia, the patient was put on the park bench position with her head fixed in Mayfield three keys head support system. Electromyographic (EMG) and auditory brainstem responses (ABRs) were utilized to monitor the cranial nerves. The tumor was totally resected via suboccipital approach. Intraoperatively, the tumor was located at the dorsal side of the brain stem in the foramen magnum of the occipital bone. The tumor was solid and soft with very scanty blood supply. The tumor was also adhering to inferior cranial nerves, posterior inferior cerebellar artery (PICA) as well as the brain stem. After careful dissection, we attained total resection of the tumor in a piece meal approach. After achieving total hemostasis, the occipital bone flap was replaced and skin closed in layers.

We observed cystic tumor samples after resection (Figure 2a). Examination of the samples at the pathology department confirmed scaly epithelium which was consistent with the diagnosis of dermoid cyst (Figure 2b). Postoperative MRI revealed total resection of the tumor (Figure 3a-b). Also, the postoperative cause was uneventful with no neurological deficits and the patient was discharged home a week after the operation. Two years follow-up revealed no recurrence of the lesion and no neurological deficits.

DISCUSSION

The wall of intracranial dermoid cyst is often depicted with thick, stratified squamous epithelium capsule that encloses dermal elements like sebaceous glands, sweat glands as well as hair follicles (3-8, 13). In almost all the cases reported in literature, the features above, most specially the presence of hair follicles were detected during histopathology evaluation (3, 5). Contrarily, we observed visible hair immediately we located the lesion intraoperatively. These lesions are frequently observed in patients between the third and fifth decades of life with no preferential sex dominance (3, 8, 14). Our case was in the third decade and a female.

Intracranial dermoid cysts are normally seen along the midline (3-8). Their midline location supports the theory of ectopic ectoderm unsuitably sequestered in the neural tube during closure between the third and fifth weeks of gestation (3-8). The lesion has a congenital origin in our patient. Nonetheless, iatrogenic dermoid cysts have been observed after procedures that inoculates epithelium into areas closer to neural tissue such as lumbar puncture or percutaneous aspiration of subdural hematoma (3, 15).

The symptomatology of intracranial dermoid depends on the location of the lesions (3, 4, 6-8). Most often patients present with headaches, seizures as well as symptoms of increased intracranial pressure (3, 4, 6-8). Nevertheless, patients frequently remain asymptomatic until spontaneous rupture of the cyst, or until the mass

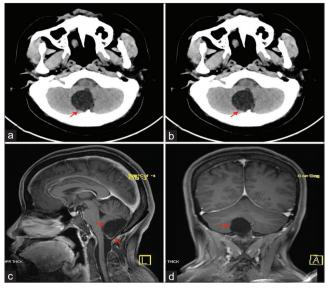


Figure 1. Preoperative MRIs. a: Is a preoperative CT scan showing a mass at occipital cistern consistent with a cystic lesion. red arrow = tumor. b-c: Are preoperative MRIs showing irregular lesions in the posterior part of the medulla oblongata with enhanced encapsulated edges. red arrow = tumor

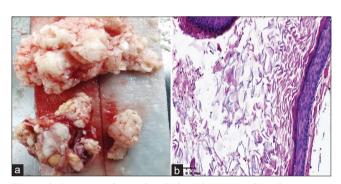


Figure 2. Intraoperative and pathology images. a: is an image of the resected dermoid cyst. b: is a histopathology imaging confirming a dermoid cyst

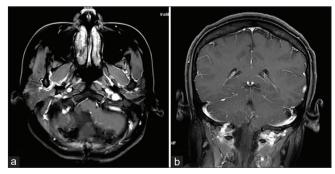


Figure 3. Postoperative MRIs. a-b: Are postoperative MRIs showing total resection of the lesion

grows up to about 3 cm in size (3, 8, 14). Our patient presented with headaches and dizziness with no nausea, vomiting or fever. Traumatic rupture of intracranial dermoid cysts often manifests as acute headache or meningismus (7, 14, 16, 17). Furthermore, ruptured posterior fossa dermoid cyst manifesting as hydrocephalus has also been reported (2, 11). Epidermoid cyst, arachnoid cyst, and

cystic craniopharyngiomas are the differential diagnosis of intracranial dermoid cysts (2, 3).

Intracranial dermoid cysts characteristically appear as non-enhancing hypodense lesions on CT scan because they are predominantly fat-filled capsules (3, 6-8). Also, punctate hyperintensities may be seen which are characteristic features of calcified component of the cyst (3, 6, 8, 18, 19). In our case, CT scan revealed a masse at occipital cistern which was consistent with a cystic lesion. However, these lesions are seen as hyperintense on T1-weighted MRI sequences with variable signals on T2- weighted sequences mainly at the solid components of the cyst (3, 5, 6, 12). Also, heterogeneous signal intensities on T2-weighted MRI varying from hypo- to hyperintense as a result of variable densities of the fatty components within the cyst have been observed (3, 8, 19). In our case, MRI revealed irregular lesions in the posterior part of the medulla oblongata with enhanced edges signifying calcifications.

In cases of ruptured cyst, the cystic contents are seen as hyperintensity on both T1- and T2-weighted MRI with the "tell-tale sign" of fat-like droplets in the subarachnoid space or ventricular system (3, 6, 8). The most key potential radiological differential diagnosis of intracranial dermoid cyst includes arachnoid cyst, cholesterol granulomas and epidermoid cyst (3, 19, 20). Heterogeneous appearance of dermoid cysts often differentiates them from arachnoid cysts on imaging (3, 19). Also, cholesterol granulomas frequently display higher inflammatory response as compared to dermoid cysts (3, 19). Nevertheless, epidermoid cysts often have a more variable location as compared to dermoid cyst because they are usually non-midline (2, 3, 15, 18).

The gold-standard treatment modality for intracranial dermoid cyst is Surgery (3-8, 12). Surgical approach is often dependent on the location of the cyst (3). Posterior fossa dermoid cysts are often resected via a suboccipital craniotomy (3, 20, 21). In case, the tumor was totally resected via the suboccipital approach. In very complex cases, total resection may not be possible when the capsule is adherent to neuro-vascular structures or the brainstem. Thus, we advocate that, EMG and ABRs should often be used to monitor the cranial nerves during resection.

We also advocate that, the goal in surgical decision-making should be safe and total resection while monitoring the cranial nerves. Rare incidence of squamous cell carcinoma in retained remnants of a dermoid cystic tumor wall after resection have been reported (18, 22, 23). Although recurrence of the lesion is rare but is more common when total resection is not attained (7). Pathological examination of specimen in case showed hair fragments as well as scaly epithelium within the specimen, consistent with a dermoid cyst as initially reported (3).

CONCLUSION

Congenital intracranial dermoid cyst in the posterior fossa is very rare lesion. We advocate that, the goal in surgical decision-making should be safe and total resection while monitoring the cranial nerves with EMG and ABRs.

Abbreviations List

Auditory brainstem responses = ABRs, Computer tomographic = CT, Computer tomographic angiograph = CTA, Electromyographic = EMG, Magnetic resonance imaging = MRI, Posterior inferior cerebellar artery = PICA.

DECLARATION

Ethics Approval and Consent to Participate

This case was reported or written in accordance to ethical committee of West China Hospital criteria for reporting or writing case reports. The patient and her relatives were informed about our intension to involve her in a case study and they agreed to partake in the study.

Consent for Publication

The patient and her relatives were dually informed about our intention to publish her case and they fully concerted to the use of these documents. Written informed consent was obtained from the patient. A written concern for publication was signed by patient. The hospital also concerted to the use of her informations for publication.

Availability of Data and Materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing Interests

All the authors have no competing interest to disclose.

Author Contributions

LL, SAR, ZL and YZ contributed toward conceptualization and data analysis, SAR performed the literature search and drafting of the paper. All authors critically revised the paper and agree to be accountable for all aspects of the work. All authors have read and approved the manuscript.

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