Deciphering a Rare Congenital Occipital Mass: A Unique Case Presentation

Sanjamjot Singh[1] Sifatjot Singh[2] Manpreet Kaur[3] Harpinder Singh Wealthy[4]

[1] Department of General Surgery, All India Institute of Medical Sciences (AIIMS) Jodhpur, Jodhpur, Rajasthan, India
[2] Department of General Surgery, Post Graduate Institute of Medical Education and Research (PGI) Chandigarh, Chandigarh, India
[3] Department of Anatomy, Muzaffarnagar Medical College, Muzaffarnagar, Uttar Pradesh, India
[4] Pediatric Surgery, Wealthy Pediatric Surgery Center, Patiala, Punjab, India

Address for correspondence H. S. Wealthy, MS, MCh, Wealthy Pediatric Surgery Center, Patiala, Punjab 147001, India (e-mail: hswealthy@yahoo.com).

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Abstract

Congenital occipital masses in neonates and infants are rare and encompass a wide range of differential diagnoses, including encephaloceles, dermoid cysts, teratomas, and other developmental anomalies. Dermoid cysts, a benign entity, arise from ectodermal sequestration during embryogenesis and can present as soft, cystic, nontender swellings at birth. Early recognition and appropriate management are essential to prevent complications and ensure favorable outcomes. We report a case of a 3-month-old male infant who presented with a soft, cystic, nontender swelling in the occipital region since birth. The mass was brilliantly translucent on clinical examination, and computed tomography imaging revealed a well-encapsulated hypodense cystic lesion with no evidence of intracranial extension or postcontrast enhancement. The child underwent complete surgical excision under general anesthesia, and intraoperative findings confirmed the lesion was extracranial, with no bony defects or neural communication. The postoperative course was uneventful, and the patient was discharged on the second postoperative day without complications. This case highlights the importance of considering congenital dermoid cysts in the differential diagnosis of occipital masses in early infancy. Accurate imaging, early surgical intervention, and histopathological confirmation are crucial for definitive management. A multidisciplinary approach ensures optimal outcomes and minimizes the risk of recurrence or complications.

Keywords

- congenital occipital mass
- dermoid cyst
- neonatal swelling
- occipital encephalocele
- pediatric neurosurgery
- case report

Introduction

Congenital occipital swellings are uncommon clinical findings in neonates and infants, presenting significant diagnostic challenges due to the broad spectrum of potential underlying conditions.[1] These swellings can arise from various congenital anomalies, including neural tube defects, vascular malformations, cystic lesions, and neoplastic growths, making early recognition and accurate diagnosis essential for appropriate management.[2] While some occipital masses are benign and self-limiting, others may have serious neurological, cosmetic, or even life-threatening implications if not identified and treated promptly.[3]

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Occipital masses in infants can be classified into dermal, neural, vascular, and neoplastic origins, with presentations varying from simple dermoid and epidermoid cysts to complex anomalies such as encephaloceles, teratomas, lipomas, and hemangiomas.[4,5] Among these, encephaloceles, resulting from defective neural tube closure leading to herniation of intracranial contents, represent a particularly significant concern due to their association with the central nervous system involvement.[6] Other differential diagnoses include lymphatic malformations, meningoceles, gliomas, and congenital fibromas, necessitating meticulous clinical and radiological evaluation for definitive diagnosis.[1]

Advancements in imaging modalities, such as ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI), have greatly enhanced the ability to characterize these lesions and determine their origin, extent, and potential involvement of adjacent structures.[2] Clinical examination, combined with a detailed patient history and radiological assessment, plays a pivotal role in guiding the diagnostic and therapeutic approach.[7]

In this report, we present a rare case of a congenital occipital mass in a 3-month-old child, detailing the clinical presentation, imaging findings, differential diagnoses, and subsequent management. Congenital dermoid cysts develop during embryonic fusion when ectodermal tissue becomes trapped along the fusion lines.[2] Dermoid cysts of the head are rare lesions comprising epidermal and mesodermal elements, with occipital localization being extremely uncommon.[4] Only a few cases of dermoid cysts in the posterior scalp have been reported, and a bilateral, synchronous presentation in this location has not been previously documented in the literature.[1]

This case underscores the importance of a multidisciplinary approach involving pediatricians, neurosurgeons, and radiologists in evaluating and treating congenital occipital swellings. Early diagnosis and timely intervention are crucial for preventing complications, optimizing neurological outcomes, and ensuring a better quality of life for affected infants.

Case Report

A 3-month-old male infant presented with a swelling in the occipital region that had been present since birth (►**Fig. 1A** and **B**). The swelling was soft, cystic, nontender, and brilliantly translucent upon clinical examination (►**Fig. 2A** and **B**). There were no signs of ulceration, discharge, or associated neurological deficits.

Radiological evaluation was conducted using CT scanning, which revealed a well-defined hypodense cystic lesion measuring between −87 and +24 Hounsfield units (HU), with an average of +3.2 HU. The lesion was encapsulated by a hypodense capsule with no postcontrast enhancement, suggesting a benign nature. Importantly, no underlying bony defect or communication with intracranial structures was identified, effectively ruling out neural tube defects such as encephalocele.

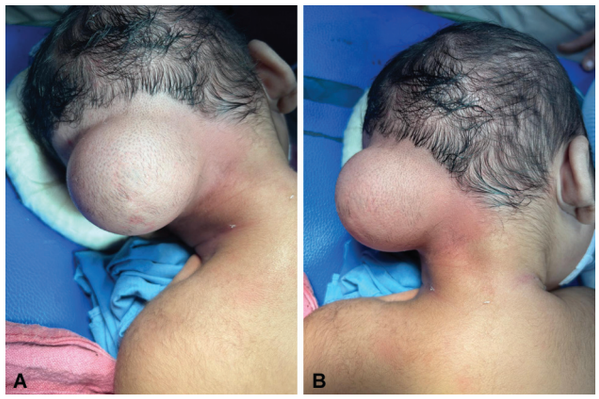

Fig. 1 (A and B) Preoperative photograph showing the occipital lesions.

The patient underwent surgical excision under general anesthesia, during which complete removal of the swelling was successfully achieved (►**Fig. 3A** and **B**). Intraoperative findings confirmed that the lesion was well-encapsulated, free from any intracranial extension, and had no communication through the occipital bone. The excised specimen was sent for histopathological examination (HPE), which confirmed the diagnosis of a congenital occipital dermoid cyst.

The postoperative period was uneventful, and the patient was discharged on the second postoperative day with no complications. Follow-up evaluations showed no recurrence or residual neurological deficits, affirming the successful surgical outcome (►**Fig. 4**). This case highlights the importance of early diagnosis and surgical management of congenital occipital dermoid cysts to prevent potential complications and ensure optimal patient outcomes.

Discussion

Congenital occipital dermoid cysts are rare benign lesions that arise from ectodermal sequestration during embryogenesis.[8] These cysts typically present at birth or in early infancy as soft, cystic, nontender masses with well-defined margins. Unlike other congenital occipital swellings, such as

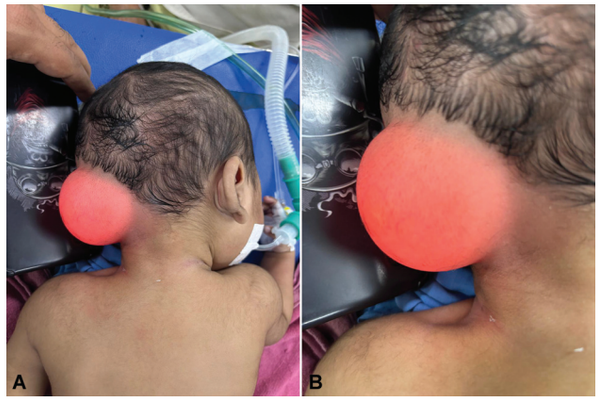

Fig. 2 (A and B) Lesion showing translucency.

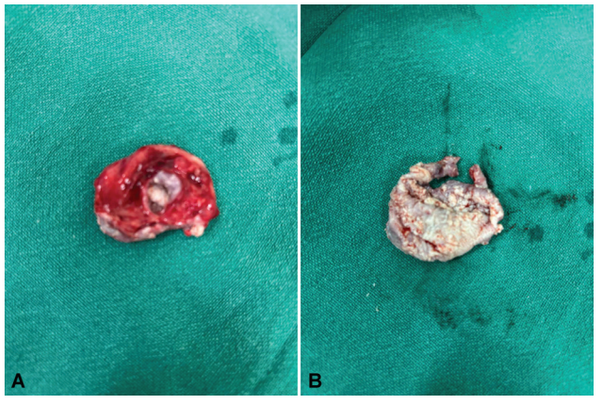

Fig. 3 (A and B) Excised lesion.

Unlike encephaloceles, dermoid cysts are usually extracranial and lack communication with intracranial structures, making them a distinct clinical entity.[9]

Dermoid cysts commonly contain keratin, sebaceous material, and epithelial debris, and their slow-growing nature allows them to remain asymptomatic for extended periods. However, complications such as infection, rupture, or secondary inflammation can arise if left untreated. The translucent nature of the swelling, as observed in this case, is a key clinical feature that aids in distinguishing dermoid cysts from other solid or vascular lesions.[10]

Radiological imaging plays a crucial role in diagnosis. CT scans are highly effective in evaluating the density of the lesion, its encapsulated nature, and any possible intracranial extension. In this case, the cyst displayed hypodense characteristics with no postcontrast enhancement, and there was no bony defect or neural communication, ruling out differential diagnoses such as encephalocele or teratoma. MRI could provide additional detail, particularly regarding soft tissue involvement, but was not necessary given the definitive findings on CT.[11]

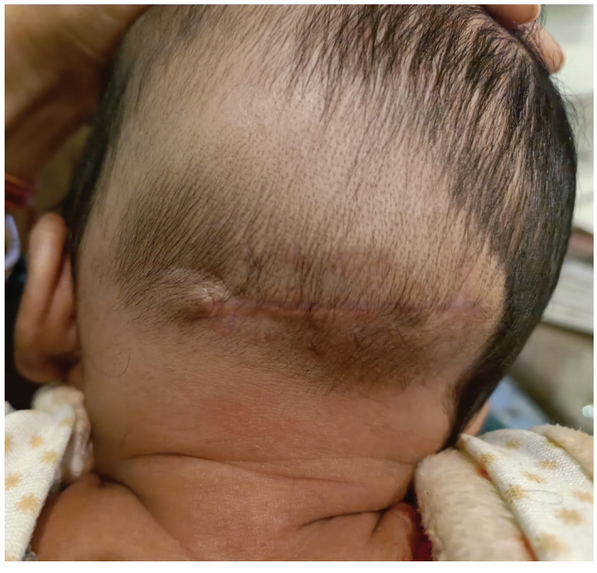

Fig. 4 Follow-up photograph showing completely healed site.

The differential diagnosis for this lesion includes encephalocele—a neural tube defect in which intracranial contents herniate through a skull defect; occipital meningocele—a type of encephalocele containing only meninges, distinguished from dermoid cysts by its connection to the subarachnoid space and potential cerebrospinal fluid leakage; epidermoid cyst—similar to dermoid cysts but lacks adnexal structures such as sebaceous and sweat glands; teratoma—a germ cell tumor containing elements from all three germ layers, often exhibiting calcifications and solid components on imaging; and hemangioma or lymphangioma—vascular malformations that present as compressible, soft tissue masses, often showing enhancement on imaging.[12,13]

The preferred treatment for congenital dermoid cysts is complete surgical excision, as was successfully performed in this case.[14] The surgical approach should ensure total removal of the cyst wall to prevent recurrence. Intraoperative findings confirmed the lesion's extracranial localization, and the absence of bony defects minimized the risk of complications.[15] HPE is essential for confirming the diagnosis, as was done in this case, demonstrating characteristic dermoid cyst histology.

The prognosis for patients undergoing early surgical excision is excellent, with a very low recurrence rate. Postoperative recovery is typically uneventful, as observed in this patient, who was discharged on the second postoperative day without any complications. This case highlights the importance of early recognition, accurate imaging, and timely surgical intervention in preventing potential complications associated with congenital occipital dermoid cysts.

Conclusion

In conclusion, congenital occipital dermoid cysts should be considered in the differential diagnosis of neonatal occipital swellings. A multidisciplinary approach involving pediatricians, radiologists, and surgeons is essential for accurate diagnosis and optimal management, ensuring favorable clinical outcomes and minimizing the risk of recurrence.

Conflict of Interest
None declared.

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