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Multiloculated Cervical Lymphangioma Associated With Periventricular Leukomalacia In A 4-Year-Old Child: A Case Report

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Abstract

Background: *Congenital cervical masses in children, such as lymphangiomas, pose diagnostic and treatment challenges due to their anatomical complexity and diverse presentations. Commonly found in the head and neck, these benign malformations rarely coexist with neurological conditions like periventricular leukomalacia (PVL), a white matter brain injury linked to perinatal hypoxia, further complicating clinical assessment and management.* ***Case Presentation:*** *This report describes a 4-year-old child with a chronic, non-tender swelling on the left side of the neck. MRI imaging revealed a multiloculated, T2 hyperintense lesion with internal septations in the left submandibular region, consistent with a lymphangioma. A secondary, similar lesion was found at the left thoracic inlet. Concurrent MRI brain findings demonstrated bilateral periventricular hyperintensities in the posterior corona radiata, confirming PVL. The child had a history of birth hypoxia and exhibited global developmental delays.* ***Diagnostic Assessment:*** *The diagnosis was supported by fine needle aspiration cytology (FNAC), which confirmed the lesion's benign cystic nature. The imaging and clinical findings effectively ruled out alternative diagnoses such as branchial cleft cysts due to the lesion's multiloculated nature and anatomical spread.* ***Management and Outcome:*** *Initial management included close airway surveillance due to the lesion's proximity to the pharyngeal space. Long-term care planning emphasized surgical or sclerotherapy options for the lymphangioma, along with ongoing neurologic monitoring and developmental interventions for PVL. A multidisciplinary team approach involving ENT, neurology, radiology, and pediatric surgery was employed.* ***Conclusion:*** *This case highlights the importance of MRI in diagnosing complex congenital cervical masses and associated neurological conditions. It underscores the need for a coordinated, multidisciplinary management strategy to address both the structural and neurodevelopmental needs of pediatric patients with dual pathologies.*

Keywords: *Lymphangioma, Periventricular Leukomalacia, Pediatric Neck Mass, Hypoxic-Ischemic Encephalopathy, MRI Diagnosis, Cystic Hygroma, Developmental Delay*

1. Introduction

Congenital neck masses in pediatric patients pose a diagnostic challenge due to their overlapping clinical and radiological features [1]. These anomalies often originate from developmental disturbances during embryogenesis, particularly involving the branchial apparatus, lymphatic system, and thyroglossal duct [2]. Their presentation may range from asymptomatic swellings to life-threatening airway obstructions depending on location, size, and involvement of adjacent structures [3]. Among these, lymphangiomas (also referred to as cystic hygromas) are benign congenital malformations of the lymphatic system, most frequently presenting in the head and neck region [4]. They typically manifest within the first two years of life and are characterized by painless, soft, multiloculated cystic swellings. Their growth is often slow but can accelerate during infections or trauma, and due to their infiltrative nature, complete surgical excision can be challenging [5].

Branchial cleft cysts, particularly second and third cleft derivatives, also contribute to congenital neck masses and are generally identified along the anterior border of the sternocleidomastoid muscle [6]. While these are usually unilocular and well-circumscribed, they can occasionally mimic the appearance of lymphangiomas on imaging. The accurate differentiation between these entities is essential for planning appropriate management strategies, as their treatment modalities and prognosis differ significantly. Magnetic Resonance Imaging (MRI) plays a pivotal role in the anatomical delineation of these lesions, evaluating their extent, internal architecture, and potential effects on nearby vital structures [7].

In rare scenarios, congenital neck masses may coincide with other systemic or neurological abnormalities, as seen in the present case [8]. The current report discusses a 4-year-old child presenting with a left-sided cervical swelling diagnosed as lymphangioma, who also demonstrated signs of periventricular leukomalacia (PVL) on brain MRI — a condition often linked with perinatal hypoxic-ischemic encephalopathy (HIE) [9]. PVL is one of the most common causes of cerebral palsy and developmental delays in children and represents white matter injury of the immature brain [10]. The present case study underscores the significance of a multidisciplinary approach, integrating radiological, clinical, pathological, and neurological evaluations to provide a comprehensive diagnosis and management plan in pediatric patients with complex congenital conditions.

2. Case description

The patient is a 4-year-old child who presented with a left-sided neck swelling. Details regarding the gender and specific geographic location were not provided in the case documentation. The child has a notable birth history marked by significant hypoxia at the time of delivery. During infancy, delays in achieving developmental milestones were observed, although no specific timeline was documented. The global nature of the developmental delay is implied and is likely attributable to perinatal asphyxia, as supported by neuroimaging findings indicative of hypoxic brain injury.

3. Materials and Methods

3.1 Study Design and Setting

This case-based observational analysis was conducted at a tertiary pediatric healthcare facility equipped with multidisciplinary services including otolaryngology (ENT), neurology, radiology, and pediatric surgery. The study received approval from the Institutional Ethics Committee, and informed consent was obtained from the patient's legal guardians.

3.2 Patient Management Protocol

Upon diagnosis, initial management focused on airway surveillance, given the anatomical proximity of the lymphangioma to the pharyngeal and upper airway structures. The patient underwent continuous clinical and radiological monitoring using MRI and ultrasonography to assess lesion size, anatomical relationships, and potential airway compromise.

3.3 Therapeutic Planning

A multidisciplinary team (MDT) approach was employed to determine optimal therapeutic strategies. Weekly MDT meetings were conducted involving consultants from ENT, pediatric surgery, neurology, and radiology to assess progress and adapt the management plan accordingly.

3.4 Surgical and Sclerotherapy Considerations

Based on lesion characteristics (e.g., size, locularity, and anatomical extent), the patient was evaluated for possible surgical excision or sclerotherapy using agents such as bleomycin or OK-432. Pre-procedural imaging guided decision-making.

3.5 Neurologic Monitoring and Developmental Intervention

The patient was also diagnosed with periventricular leukomalacia (PVL). Ongoing neurologic evaluations were performed using cranial MRI, electroencephalography (EEG), and clinical assessments. The patient was enrolled in a neurodevelopmental therapy program, including physical, occupational, and speech therapy, coordinated by pediatric neurology.

3.6 Outcome Assessment

Clinical follow-up was conducted over a defined observation period to comprehensively evaluate the patient's response to the multidisciplinary management approach. Particular attention was given to monitoring airway status and respiratory function due to the lesion's proximity to the pharyngeal space, with regular clinical assessments and imaging to detect any signs of obstruction or compromise. The therapeutic response was assessed by evaluating the reduction in lesion size or complete resolution through serial imaging studies such as MRI and

ultrasonography. In parallel, neurologic development was closely monitored using standardized developmental milestone checklists and neurologic scoring systems to detect any deficits associated with periventricular leukomalacia (PVL). These evaluations provided critical insights into the effectiveness of both surgical and supportive interventions and guided necessary modifications in the long-term care plan.

4. Observations

4.1 Clinical Presentation

The chief complaint in this case is a gradual, non-tender swelling located on the left side of the neck. Although the exact duration of the swelling is not specified, it is presumed to be chronic based on the imaging characteristics and clinical presentation. Notably, the child does not report any associated symptoms such as pain, difficulty in swallowing (dysphagia), breathing issues, hoarseness of voice, or any history indicative of an acute infection.

4.2MRI Neck Findings

Imaging of the neck revealed a primary lesion in the left submandibular region characterized by T1 hypointensity and T2 hyperintensity. The lesion appeared well-defined and multiloculated with both thin and thick internal septations. It measured approximately 3.8 x 1.8 cm and was situated in the region corresponding to the second and third branchial clefts. The lesion extended adjacent to the pharynx and posteriorly into the retropharyngeal space, with an enveloping phenomenon noted around the supraglottic area. Importantly, there was no involvement of the posterior triangle. A secondary lesion was also identified at the thoracic inlet on the left side. This lesion was T2 hyperintense, measured approximately 2.1 x 1.0 cm, and displayed similar internal septated characteristics to the primary lesion.

Other anatomical structures within the imaged field were normal. The nasopharynx, oropharynx, and laryngopharynx showed normal configuration and signal intensity. The epiglottis, vallecula, and pyriform fossae were unremarkable, as were the larynx and vocal cords. There was no significant cervical lymphadenopathy. Normal flow voids were seen in the carotid arteries and internal jugular veins. The thyroid gland exhibited normal morphology of both lobes and the isthmus, with no evidence of focal lesions. The base of the skull also appeared normal.

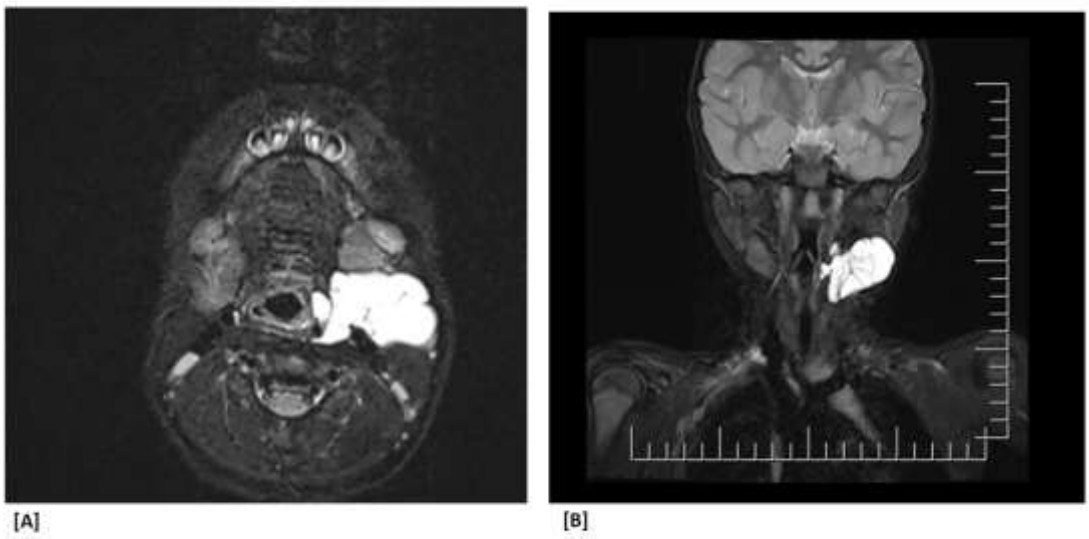

Fig. 1: Illustration [A] and [B] showing the hyperintence lesion with septa involving the region of left submandibular gland and enveloping the region of the supraglottic region posteriorly and anteriorly and anteriorly on axial and coronal images of T2WI.

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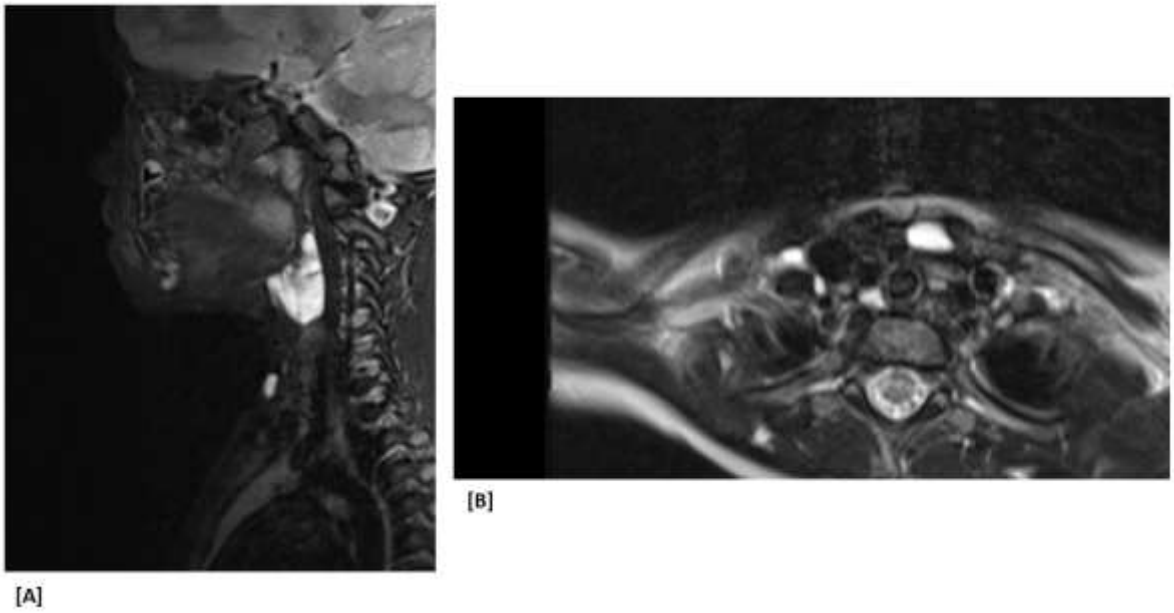

Fig. 2: Illustration [A] showing the SAG images of T2 hyperintense lesion involving the region and [B] axial images showing the second lesion at the level of thoracic inlet.

4.3 MRI Brain Findings

Neuroimaging findings revealed evidence of periventricular leukomalacia (PVL), characterized by periventricular hyperintensities noted in the posterior corona radiata bilaterally. These changes are indicative of white matter injury, most likely resulting from perinatal hypoxic-ischemic events. PVL is a known sequela of neonatal hypoxia and is frequently associated with neurodevelopmental delays and motor dysfunction in pediatric patients.

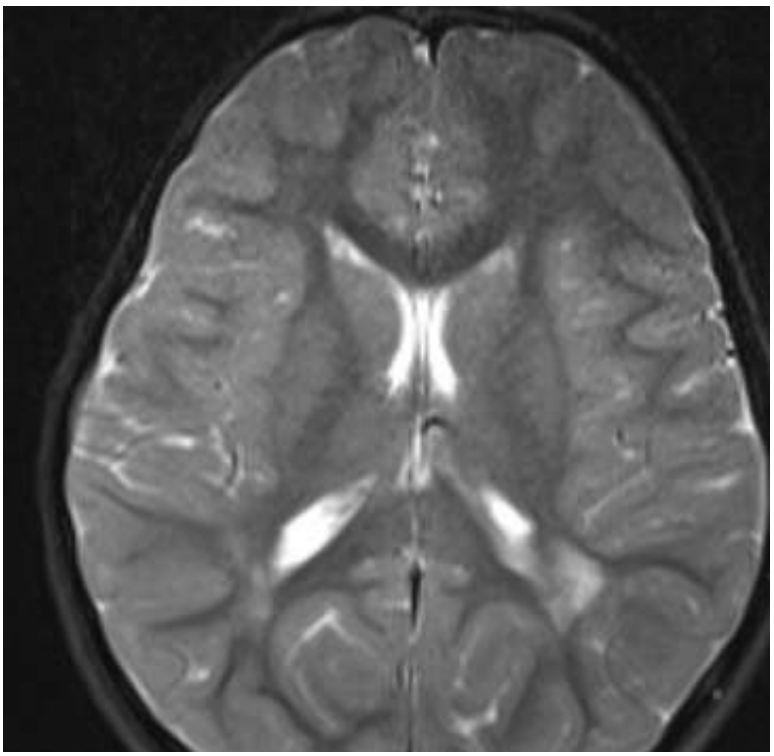

Fig 3: MRI visualized brain in the neck study showing periventricular leukomalacia (sequelae of old hypoxia).

4.4 Provisional Diagnosis

Based on the clinical presentation and radiological findings, the provisional diagnosis primarily pointed toward a lymphangioma (cystic hygroma). This was strongly supported by the lesion's multiloculated appearance, presence of both thin and thick internal septations, retropharyngeal extension, and the characteristic enveloping effect around the supraglottic structures. A branchial cleft cyst, specifically of second or third cleft origin, was considered as a differential diagnosis; however, it was deemed less likely due to the typically unilocular structure of branchial cleft cysts and the relatively limited anatomical spread they exhibit compared to the extensive, septated, and infiltrative features observed in this case.

4.5 Confirmatory Test

Fine Needle Aspiration Cytology (FNAC) was performed to further evaluate the nature of the left-sided neck swelling. The procedure involved the aspiration of fluid from the lesion, which was then subjected to cytological examination. The cytology report revealed features consistent with a benign cystic lesion. Based on the clinical context and imaging correlation, the final impression from FNAC supported the diagnosis of a probable lymphangioma located in the left cervical region.

4.6 Disease Management Plan

Short-term management focused on careful monitoring of airway patency, given the lesion's proximity to vital pharyngeal structures, to ensure there was no compromise in breathing or swallowing. Additionally, a comprehensive neurological assessment was essential to evaluate the extent of developmental delays, especially in light of the confirmed periventricular leukomalacia.

Long-term management strategies included planning for surgical excision or sclerotherapy of the lymphangioma, depending on the progression of symptoms, lesion growth, or functional impairment. Ongoing neurological follow-up was advised to address complications associated with PVL, with an emphasis on early developmental interventions such as speech therapy, physiotherapy, and occupational therapy. A multidisciplinary care team comprising a pediatric neurologist, ENT surgeon, and pediatric surgeon was recommended to provide comprehensive and coordinated care tailored to both the structural and neurological needs of the patient.

5. Discussion

Lymphangiomas are benign, congenital malformations of the lymphatic system, arising from sequestrated lymphatic tissue that fails to communicate normally with the rest of the lymphatic or venous systems during embryogenesis [1, 2]. They account for approximately 6% of all benign tumors in children and are most frequently diagnosed by the age of two [11]. While the posterior cervical triangle is the most common site, these lesions can also occur in less typical locations such as the submandibular region, retropharyngeal space, and even within the thoracic inlet, as observed in this case. Their clinical significance depends not only on their size but also on their location and relationship to vital structures, particularly in the head and neck region where they may impinge on the airway or swallowing pathway [12].

The imaging hallmark of lymphangiomas is a multiloculated cystic mass with both thin and thick septations, appearing T1 hypointense and T2 hyperintense on MRI. These features help to differentiate them from other congenital lesions such as branchial cleft cysts, which tend to be unilocular, less extensive, and more commonly restricted to the lateral aspect of the neck [13]. The enveloping effect, retropharyngeal extension, and presence of a second lesion at the thoracic inlet in this case further support the diagnosis of a lymphangioma [14].

A notable aspect of this case is the concurrent diagnosis of periventricular leukomalacia (PVL), a form of hypoxic-ischemic encephalopathy (HIE) typically seen in preterm infants or those who have suffered significant birth asphyxia [15]. PVL represents the most common form of cerebral white matter injury in neonates, often associated with cognitive impairments, spastic motor deficits, and delayed developmental milestones. MRI findings of periventricular hyperintensities in the posterior corona radiata bilaterally in this patient are classic for PVL and correlate well with the clinical history of hypoxia at birth and delayed neurodevelopment [16].

The coexistence of a congenital cervical lymphatic anomaly with perinatal brain injury makes this case particularly unique. While these conditions are not pathophysiologically linked, their combined presence in a single pediatric patient demands a holistic and multidisciplinary approach to management. Airway safety, lesion progression, and the potential for surgical or sclerosing therapy must be balanced with the ongoing neurological needs of the child,

including access to early intervention programs, neurodevelopmental therapy, and long-term surveillance for motor and cognitive delays [17].

Furthermore, this case illustrates the critical role of imaging, particularly MRI, in both identifying and characterizing congenital anomalies as well as uncovering coexisting conditions that may otherwise remain undiagnosed [18]. It also highlights the importance of early and integrated evaluation by a team comprising pediatricians, radiologists, neurologists, ENT and pediatric surgeons, and rehabilitation specialists, all of whom contribute to optimizing the long-term outcomes for such complex pediatric patients [19].

6. Clinical Significance

This case study offers critical insights into the diagnostic and therapeutic challenges in pediatric care, particularly when dealing with congenital cervical anomalies. It emphasizes the pivotal role of Magnetic Resonance Imaging (MRI) as the gold standard for evaluating congenital neck masses. MRI's high-resolution, multiplanar imaging capability allows for precise characterization of lesion morphology, including the identification of multiloculated, cystic structures typical of lymphangiomas. Moreover, its ability to delineate the lesion's anatomical extent and proximity to critical structures—such as the airway and vascular elements—renders it indispensable for both diagnosis and pre-surgical planning.

In addition, the case underscores the complementary value of Fine Needle Aspiration Cytology (FNAC). FNAC not only assists in confirming the benign nature of the lesion but also serves as a useful adjunct in distinguishing lymphangiomas from other differentials such as branchial cleft cysts or inflammatory swellings. The integration of radiological and cytological findings supports accurate diagnosis, which is essential for formulating an effective treatment strategy.

A particularly noteworthy aspect of this case is the coexistence of dual pathologies—a benign cervical lymphangioma and periventricular leukomalacia (PVL), a form of white matter brain injury often associated with perinatal hypoxia-ischemia. This dual pathology presents a unique diagnostic and management challenge: while the lymphangioma requires timely surgical intervention to prevent airway or functional compromise, the PVL necessitates long-term neurological surveillance and early developmental intervention to mitigate potential neurodevelopmental deficits. Ultimately, the case reinforces the necessity of an early, accurate, and multidisciplinary approach. Coordinated management involving radiologists, pediatricians, ENT specialists, neurologists, and pediatric surgeons is crucial not only for immediate treatment but also for ensuring favorable long-term outcomes. This integrated care model can significantly enhance the quality of life and developmental trajectory of children affected by complex congenital conditions.

7. Conclusion

The present case study highlights the importance of Magnetic Resonance Imaging (MRI) in accurately identifying congenital cervical masses, such as lymphangiomas. MRI provides high-resolution imaging, crucial for diagnosis and surgical planning. Fine Needle Aspiration Cytology (FNAC) is also valuable for confirming benign cystic lesions and distinguishing between lymphatic malformations and other entities. The case highlights the clinical complexity of dual pathologies, requiring early diagnosis, vigilant monitoring, and a multidisciplinary approach involving radiologists, pediatricians, neurologists, ENT specialists, and pediatric surgeons.

Conflict of Interest

Authors declared that there is no conflict of interest.

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Ethics Approval and Consent to Participate

All necessary consent & approval was obtained by authors.

Consent for Publication

All necessary consent for publication was obtained by authors.

Data Availability
All data generated and analyzed are included within this research article.

Author Contributions
All authors contribute significantly in this manuscript.

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