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Atypical Cystic Hygroma in the Upper Back - A Congenital Swelling at Rare Presentation

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Abstract: ***Background: Cystic hygroma is a congenital lymphatic malformation commonly occurs in the cervicofacial region, with rare occurrences in atypical sites such as the upper back. Early diagnosis and appropriate management are essential to prevent complications such as infection, hemorrhage, or recurrence. Case Presentation: We present a case of a 1-year-old male child with a progressively enlarging, painless swelling over the left upper back, noted since birth. Clinical examination revealed a soft, non-tender, fluctuant mass with brilliant transillumination. Ultrasonography identified a well-defined, multicystic lesion in the subcutaneous plane without any communication with the thoracic cavity. Intraoperatively, a single cystic swelling was found, and complete excision was performed. Histopathology confirmed the diagnosis of cystic hygroma, revealing multiple cystically dilated lymphatic spaces lined by endothelial cells with minimal lymphocytic infiltration. Conclusion: This case highlights the importance of recognizing atypical presentations of cystic hygroma to ensure timely diagnosis and optimal management. Complete surgical excision remains the preferred treatment to prevent recurrence. Raising awareness of such rare occurrences aids clinicians in better decision-making and management of congenital lymphatic malformations.***

Keywords: Cystic hygroma, Cystic lymphangioma, congenital lymphatic malformation, upper back swelling, surgical excision

1. Introduction

Cystic hygroma is a congenital lymphatic malformation arising due to abnormal sequestration of lymphatic tissue during embryogenesis. It is a benign but potentially progressive condition that commonly manifests in the cervicofacial region, accounting for approximately 75–80% of cases. Other less common locations include the axilla, mediastinum, retroperitoneum, and inguinal region.[1] Cystic hygromas of the upper back are exceedingly rare and often pose diagnostic challenges due to their atypical presentation and potential overlap with other soft tissue masses.

The pathogenesis of cystic hygroma is attributed to the failure of lymphatic sacs to establish proper communication with the venous system, leading to the formation of fluid-filled cystic spaces. These lesions are typically multiloculated, soft, and transilluminant, with variable rates of growth. While some hygromas remain stable, others may expand rapidly, causing functional impairment, secondary infections, or hemorrhage.[2,3]

Early diagnosis is crucial to prevent complications, particularly in pediatric patients, where large lesions may exert pressure on surrounding structures. Ultrasound is the preferred initial imaging modality due to its high sensitivity in detecting the cystic nature of the lesion. Magnetic resonance imaging (MRI) further aids in assessing the extent of deep tissue involvement. The definitive treatment remains complete surgical excision, aiming to minimize recurrence while preserving surrounding anatomical structures.[4,5]

This case report highlights a rare presentation of cystic hygroma in the upper back of a 1-year-old child, emphasizing the clinical, radiological, and surgical aspects. The report aims to contribute to existing literature on atypical presentations and optimal management strategies, thereby enhancing clinical decision-making in similar cases.

2. Case Presentation

Patient Information

A 1-year-old male child presented to the surgical outpatient department with a painless swelling over the left upper back. The swelling was initially noted at birth but had been gradually increasing in size over the past two months. There was no history of trauma, fever, redness, or tenderness over the swelling. The child had no difficulty in movement of the upper limb, respiratory distress, or feeding difficulties. There was no history of similar swellings in the family or any significant antenatal or perinatal complications.

The child's growth and developmental milestones were age-appropriate, and systemic examination revealed no abnormalities. The family history was non-contributory, and there was no history of consanguinity.

Clinical Examination

On inspection, a well-defined, soft, non-tender swelling measuring approximately 8 cm × 5 cm was noted over the left upper back, just above the scapular region. The overlying skin was normal, with no signs of inflammation, ulceration, or discoloration. The swelling exhibited a fluctuation-positive and brilliant transillumination-positive response (***Figure: 1***), characteristic of a cystic lesion.

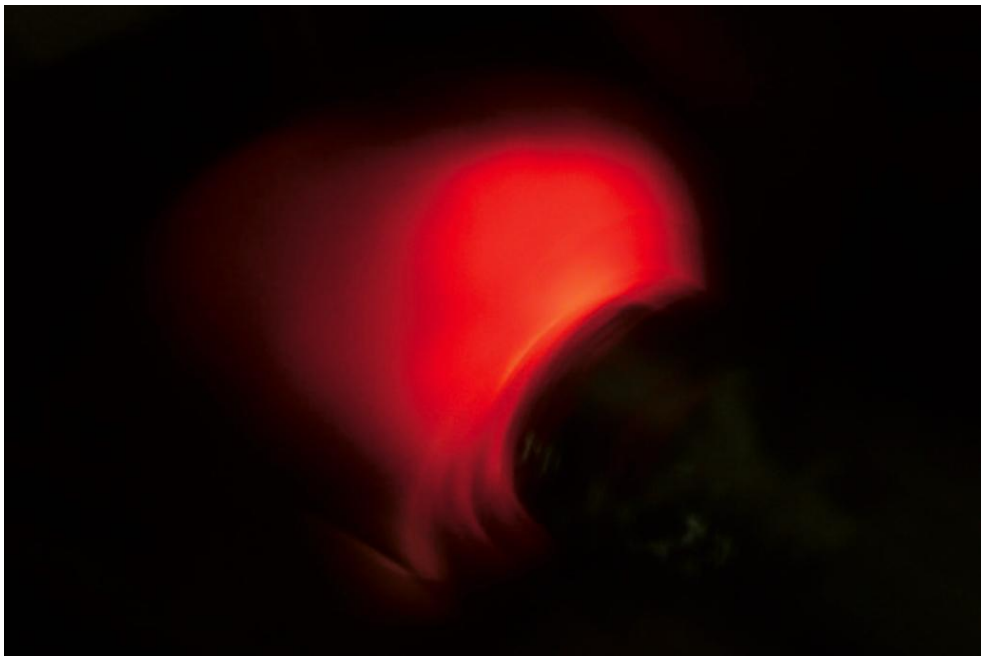

Figure 1: Brilliant transillumination positive

On palpation, the swelling was compressible, non-pulsatile, and non-adherent to underlying structures (***Figure 2***). There was no bruit or thrill, and the swelling did not change in size with respiration or straining, ruling out vascular malformations. There were no signs of regional lymphadenopathy or involvement of deeper structures.

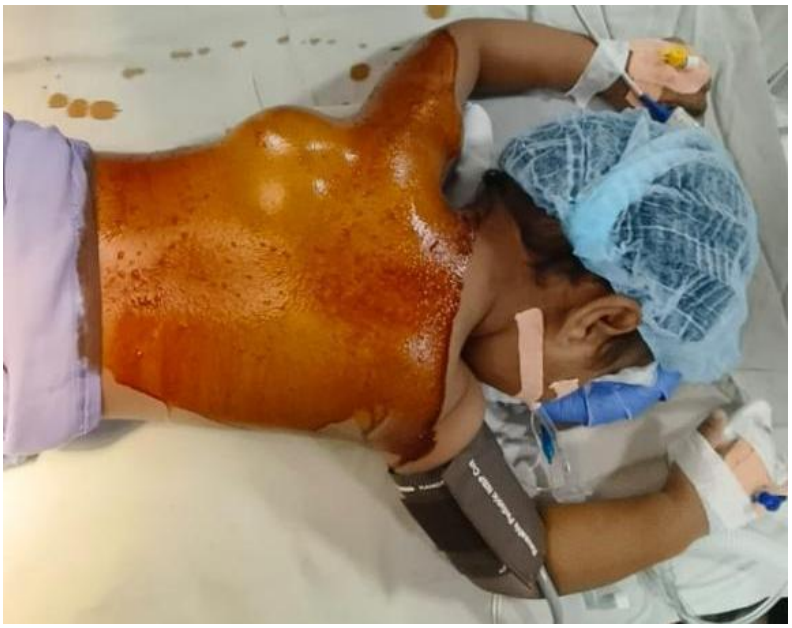

Figure 2: Pre-operative image showing swelling in left lower back

Based on these findings, a preliminary diagnosis of congenital cystic hygroma was considered, warranting further radiological evaluation to confirm the diagnosis and assess the extent of the lesion.

Diagnostic workup

To confirm the preliminary diagnosis of cystic hygroma and assess the extent of the lesion, an ultrasound evaluation was performed. Ultrasonography (USG) revealed a multiloculated anechoic cystic lesion, (***Figure 3***) measuring approximately 7.9 cm × 4.4 cm × 2.8 cm, situated in the deep subcutaneous plane of the left upper back over the scapular region. The lesion exhibited well-defined margins with multiple cystic spaces, characteristic of a lymphatic malformation. Notably, there was no evidence of internal vascularity, ruling out vascular malformations such as hemangioma. The lesion also demonstrated no communication with the underlying thoracic cavity, indicating its isolated subcutaneous nature.

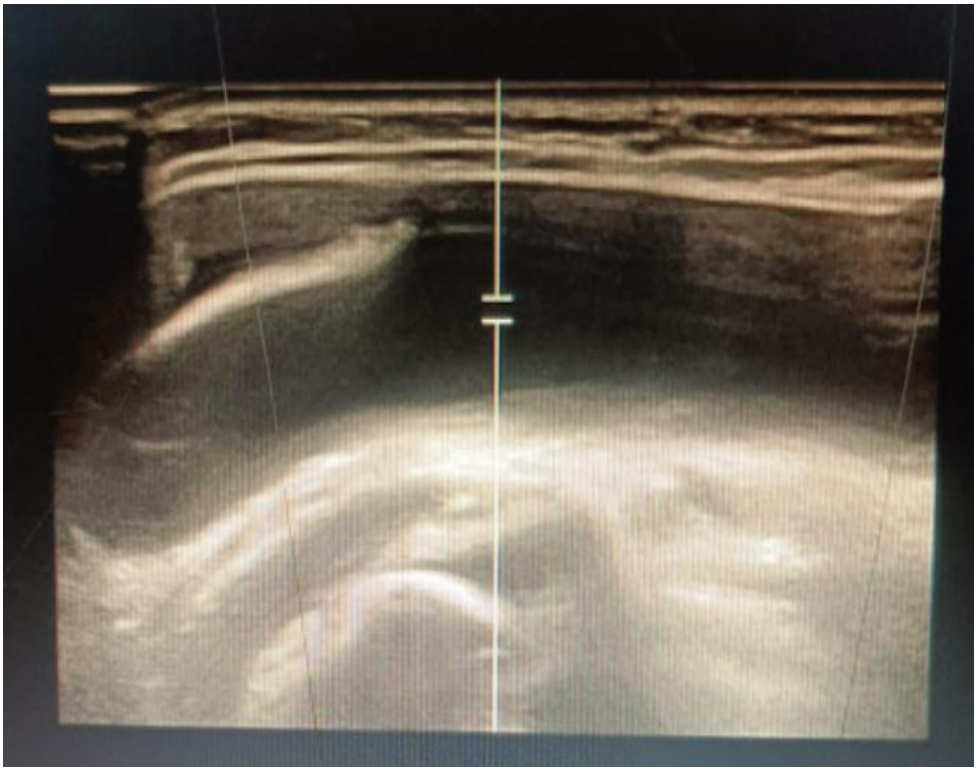

Figure 3: Ultrasound shows swelling in the subcutaneous plane

Given the radiological findings, a differential diagnosis of lipoma, dermoid cyst, and sebaceous cyst was considered; however, the classic features of a multiloculated cystic lesion with transillumination positivity strongly favored cystic hygroma. Based on clinical and radiological correlation, a definitive diagnosis of congenital cystic hygroma was established, leading to the decision for surgical excision.

Surgical Findings & Management

The patient was planned for surgical excision under general anesthesia, given the increasing size of the lesion and its potential for complications. Intraoperatively, a single cystic swelling measuring approximately 8 cm × 4 cm × 3 cm was identified in the left upper back, located in the subcutaneous plane without any connection to the thoracic cavity. The lesion was well-encapsulated, multiloculated, and cystic, with no apparent infiltration into deeper structures. (***Figure 4***)

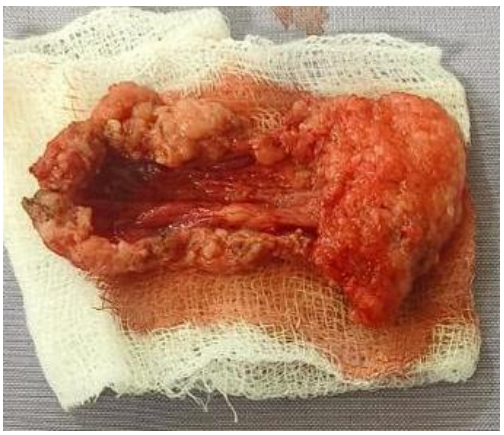

Figure 4: Post-operative image showing excised cyst

Careful blunt and sharp dissection was performed to completely excise the lesion while preserving surrounding vital structures. The cystic mass was removed intact to prevent spillage, minimizing the risk of recurrence. Meticulous hemostasis was ensured, and the surgical site was closed in layers. The postoperative period was uneventful, with no signs of infection, hematoma, or recurrence. The patient was monitored for wound healing and functional recovery, with a favorable postoperative outcome.

Histopathology

The excised specimen was subjected to histopathological examination, confirming the diagnosis of cystic hygroma. Microscopic analysis revealed multiple cystically dilated spaces, (***Figure 5***) lined by a single layer of flattened endothelial cells, consistent with a lymphatic origin. These cystic spaces were found within a fibrous stromal background, indicating a benign lymphatic malformation.

Additionally, a thin layer of smooth muscle cells was noted around the cystic spaces, with minimal lymphocytic inflammation, further supporting the diagnosis. There was no evidence of dysplasia, malignancy, or vascular invasion. The histopathological findings confirmed that the lesion was a benign congenital lymphatic malformation, reinforcing the appropriateness of surgical excision as the definitive treatment. The patient was advised regular follow-up to monitor for potential recurrence, although the prognosis remained excellent.

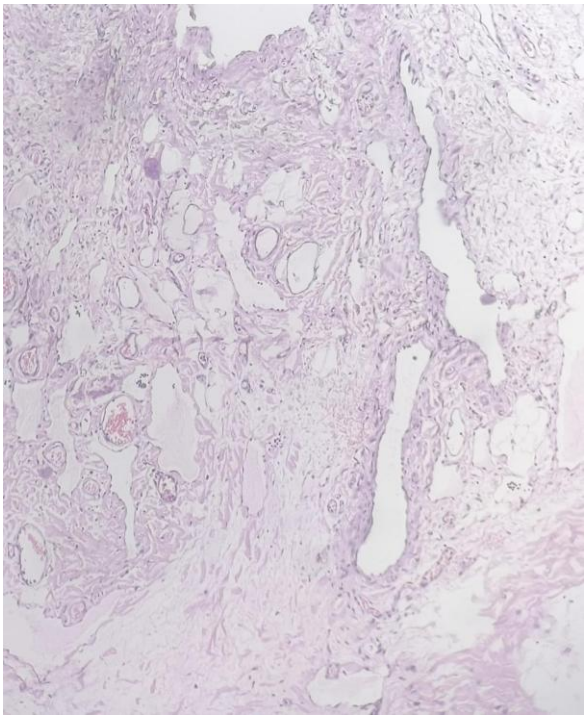

Figure 5: Histopathology: H& E staining of the cyst showing multiple cystic dilated space

3. Discussion

Cystic hygroma, also known as lymphangioma, is a rare congenital lymphatic malformation resulting from the failure of lymphatic vessels to properly communicate with the venous system during embryogenesis. It was first described by Redenbacher in 1828 and is classified into three variants: microcystic, macrocystic, and mixed types, based on cyst size and distribution. The majority of cases occur in the cervicofacial region (80%), followed by the axilla, mediastinum, groin, and tongue.[6,7] Reports of cystic hygroma in uncommon sites such as the upper back, suprasternal region, and extremities are exceedingly rare.[8–10]

A significant proportion of cystic hygromas (over 60%) manifest at birth, with 90% becoming apparent by two years of age. They are often diagnosed due to painless, progressively enlarging swelling, which can lead to compression of adjacent structures. While small lesions may remain asymptomatic, larger or deep-seated cystic hygromas can impair respiration, swallowing, or limb movement. Differential diagnoses include lipomas, sebaceous cysts, and dermoid cysts, necessitating imaging and histopathological confirmation.[11]

Cystic hygromas arise due to congenital lymphatic sequestration, where primitive lymphatic channels fail to connect with the central venous system, leading to fluid accumulation within malformed lymphatic spaces. These cystic spaces are lined by endothelial cells and supported by a fibrous stroma, with little to no smooth muscle in their walls, explaining their propensity to enlarge over time due to fluid collection and lack of structural integrity.[12,13]

Ultrasound typically reveals a multiloculated, anechoic cystic lesion, while MRI and CT scans provide further insights into extent, depth, and potential communication with surrounding structures. Surgical excision remains the gold standard for treatment, particularly in large or symptomatic lesions, with alternative modalities such as sclerotherapy used for inoperable cases.[14]

Cystic hygroma, a congenital lymphatic malformation, presents as a soft, fluctuant swelling, often necessitating differentiation from other cystic or soft tissue lesions. Key differential diagnoses include lipoma, characterized by its soft, mobile, and non-tender nature, and lack of fluctuation and transillumination. Dermoid cysts, congenital lesions containing ectodermal derivatives, are typically firm and may contain hair or other contents. Sebaceous cysts, filled with keratin and sebum, often exhibit a punctum. Branchial cleft cysts, arising from embryological remnants, are located in the lateral neck. Lymphadenopathy, characterized by enlarged lymph nodes, typically presents with firmer consistency and lacks transillumination.[15,16]

The management of cystic hygroma depends on its size, location, and associated complications. Surgical excision is the preferred approach for well-defined, accessible lesions, aiming for complete removal to minimize the risk of recurrence. For inoperable or recurrent lesions, sclerotherapy with agents like Bleomycin, OK-432, or Doxycycline may be considered. Laser therapy and radiofrequency ablation can be utilized for smaller, superficial lesions. In certain cases, observation may be an option for small, asymptomatic lesions that may spontaneously regress.[17,18]

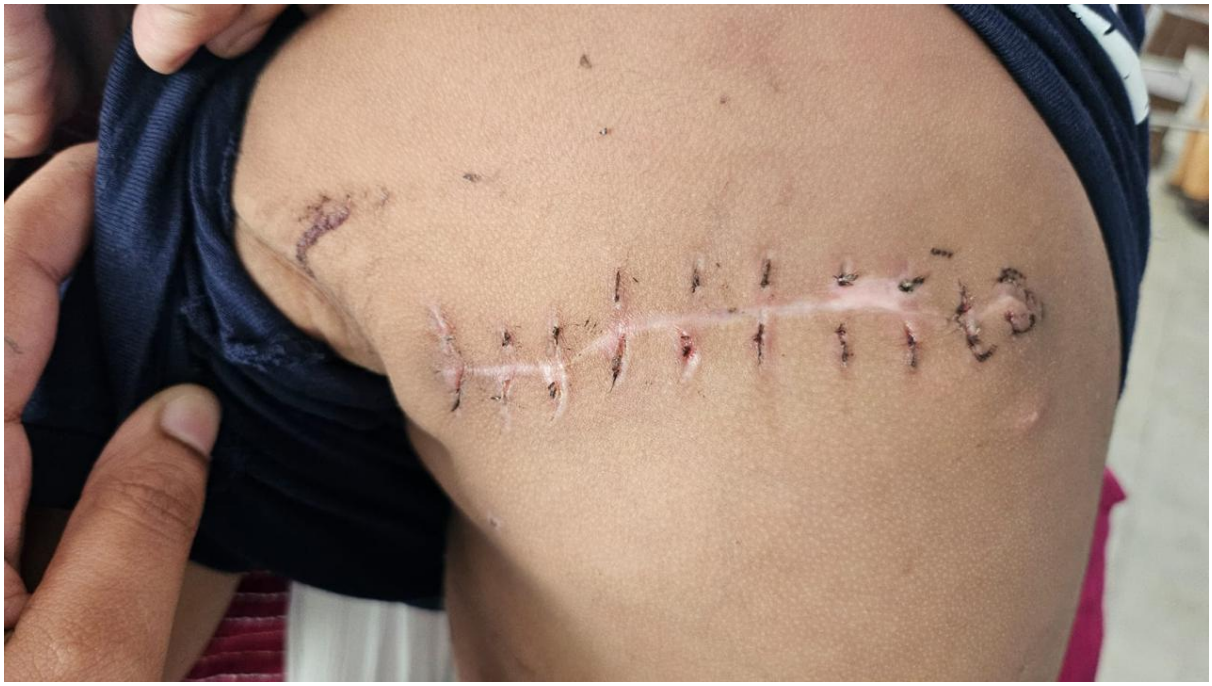

Figure 6: Follow up picture

Complications

Cystic hygroma presents several potential complications. Incomplete surgical excision can result in recurrence, necessitating further intervention. Furthermore, secondary bacterial infections can occur, leading to pain, erythema, and the formation of abscesses. Vascular involvement within the cyst may increase the risk of internal hemorrhage. Finally, large cystic hygromas located in the neck can compromise the airway, potentially causing difficulty in breathing and swallowing, necessitating prompt medical attention.

4. Conclusion

Cystic hygroma is a congenital lymphatic malformation that typically presents in the cervicofacial region. However, its occurrence in the upper back is exceedingly rare, posing a diagnostic challenge. Early recognition of such atypical presentations is crucial for ensuring timely intervention and optimal patient outcomes. A thorough clinical evaluation, complemented by imaging studies, facilitates accurate diagnosis and differentiation from other cystic or soft tissue lesions.

Complete surgical excision remains the gold standard for treatment, as it significantly reduces the risk of recurrence and associated complications such as infection and hemorrhage. Early surgical intervention prevents potential functional and cosmetic concerns, thereby improving the overall prognosis.

Raising awareness among clinicians regarding unusual presentations of cystic hygroma enhances diagnostic accuracy and aids in formulating appropriate management strategies. This case underscores the importance of vigilance in identifying rare congenital anomalies and highlights the need for further research to expand the understanding of such conditions.

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