

Clinical Insights: Three Case Reports of Neonatal Head and Neck Cystic Hygroma

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Abstract

Cystic hygroma, a congenital malformation characterized by fluid-filled cysts typically found in the head and neck region, presents unique challenges in neonatal care. This report examines three distinct cases of neonates diagnosed with head and neck cystic hygroma. The cases highlight the diverse clinical presentations, diagnostic considerations, and therapeutic interventions employed in managing this condition. By exploring these cases, we aim to contribute to the understanding of cystic hygroma in neonates, fostering improved clinical awareness and decision-making for healthcare professionals involved in neonatal care.

1. Introduction

Fetal lymphangioma stands out as one of the prevalent yet benign congenital malformations affecting the lymphatic system. Constituting approximately 25% of all benign vascular tumors in the pediatric population, this condition is commonly identified through prenatal ultrasound screenings. While lymphangiomas can manifest in various locations within the skin or subcutaneous tissue, the majority (75%) tend to impact the head and neck region, particularly in the posterior cervical triangle. The remaining 25% may be present in areas such as the axillae, mediastinum, groin, and retroperitoneum. The two main categories of lymphangiomas are deep and superficial, with the former further divided into cavernous lymphangiomas and cystic hygromas. Superficial types include lymphangioma circumscriptum and acquired lymphangioma, also referred to as lymphangiectasia. The predominant form, cystic hygroma, comprises single or multiple cystic lesions [1]. These cysts are categorized based on size into macrocystic (diameter >2 cm), microcystic (diameter < 2 cm), and mixed (cysts of variable sizes). The content of the cysts can be chylous or serous. The development of cystic lymphangiomas is attributed to abnormalities in the lymphatic system's embryonic development or defects in the connection between the lymphatic and venous systems. These disruptions during embryogenesis lead to impaired

lymphatic drainage, resulting in the dilation of lymphatic tissue and the formation of cystic lesions.

2. Case Presentations

We present three cases of cystic hygroma observed at our institution over 4 months. All three cases were categorized as macrocystic, with two of them identified as giant cystic hygromas. Each newborn was admitted to our neonatal intensive care unit within the first week of life due to respiratory distress, necessitating intubation for airway protection. Despite the presence of the cystic mass, the physical examination of all three newborns revealed no additional abnormal features. Notably, all three infants were Syrian refugees without adequate prenatal care.

In the cases of Baby 1 and Baby 2, both presented with a neck mass and underwent urgent aspiration to temporarily reduce the size of the cystic hygroma. Replacement therapy was initiated while awaiting surgical intervention. Unfortunately, both infants experienced cardiac arrest a few hours after the aspiration procedure and attempts at resuscitation were unsuccessful. Baby 3 had an oral mass requiring recurrent aspirations for three weeks [2-6]. However, this infant, also a Syrian refugee, was lost to follow-up, and there is no available information on the current medical status.

Clinical diagnosis for all three cases was established based on the characteristic cystic lesion that transilluminates upon examination. Ultrasound imaging performed at the time of aspiration revealed multicystic lesions with internal septations, supporting the clinical diagnosis. These cases underscore the challenges in managing cystic hygroma, particularly in the context of limited prenatal care for Syrian refugee populations. Early intervention and close follow-up are crucial for optimizing outcomes in such cases.

3. Discussion

Cystic hygromas, although capable of occurring anywhere in the body, are most observed as isolated lesions on the neck, axillae, or groin. Typically identified during the neonatal period or early infancy, these lesions often present as painless, soft, and visible masses that transilluminate. Complications may arise, and patients can develop symptoms such as tenderness, warmth, and abscess formation if the lesion becomes infected. Chromosomal abnormalities, especially Turner syndrome, are highly associated with congenital lymphangiomas, prompting the need for routine karyotype evaluations. Unfortunately, financial constraints hindered the completion of karyotype assessments for our patients [7].



Figure 1: Baby 3, born term, presented for respiratory distress secondary to cystic hygroma of the mouth. He required recurrent aspiration of the hygroma.

While cystic hygromas are generally benign, complications can manifest. Infections, either primary or secondary to nearby microbial spread, can lead to abscess formation and necessitate antibiotic treatment along with drainage if required. Enlargement of the lesion may occur during systemic viral or bacterial infections, and spontaneous hemorrhage into the cyst is a reported complication that demands urgent surgical intervention. Additionally, respiratory compromise and dysphagia may arise due to compression from a neck mass, with severe respiratory distress potentially requiring urgent interventions such as tracheostomy Figure 1.

All three of our cases presented with a mass in the head and neck region, leading to respiratory compromise and necessitating intubation within the first week of life. The challenges faced in managing these cases were exacerbated by the absence of karyotype evaluations due to financial constraints. This underscores the importance of addressing financial barriers to ensure comprehensive diagnostic evaluations and appropriate management strategies for neonates with cystic hygroma.

Diagnosis of cystic hygromas can be achieved through physical examination at birth, during infancy, or via prenatal ultrasonography. Ultrasonography reveals multicystic lesions with internal septations and no blood flow on Doppler imaging. Additional imaging techniques like CT scans and MRI are crucial for surgical planning, offering detailed visualization of the lesion and its relationship with adjacent

nerves and vessels. MRI is highly reliable, displaying the full extent of complex lesions with hyperintensity on T2 sequences due to high fluid content and demonstrating fluid-filled levels reflecting the layering of proteins and blood. Histologically, cystic hygromas exhibit large irregular vascular spaces lined with a single layer of endothelial cells, surrounded by fibroblastic, collagenous, or fatty stroma. The cysts are filled with eosinophilic and proteinaceous fluid [8].

Although cystic hygromas can remain asymptomatic, treatment is necessary for complications. Complete surgical excision is the preferred method, but it carries risks, including recurrence in 20% of cases, wound infection, scar formation, and incomplete resection. Aspiration can be performed temporarily, but recurrence is a concern. Sclerotherapy with bleomycin or OK432 has shown promise in recent years. Other modalities like radiation, laser excision, radio-frequency ablation, and cauterization yield variable results. In the presented cases, aspiration was associated with recurrence, and no surgical excisions were performed.

4. Conclusion

Cystic hygromas pose challenges, especially when presenting early in life, and larger lesions are linked to increased morbidity and mortality. Long-term follow-up data are limited, necessitating further prospective studies to compare the effectiveness of different treatment modalities. The presented cases, involving Syrian refugee infants, prompt questions about potential environmental factors related to this malformation, underscoring the need for future research.

5. Acknowledgments

None.

6. Conflicts of Interest

None.

7. References

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