International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Neonatal Mediastinal Cystic Hygroma: Case Report

Faten Alawaysheh (M. D)[1], Hashem E. Aqrabawi (M. D)[2], Alaeddin Alali (M. D)[3], Abdallah Almhairat (M. D)[4], Ghadeer Alsawalgeh (M. D)[5]

Abstract: ***A cystic hygroma is a fluid - filled sac that results from a blockage in the lymphatic system. It is most commonly located in the neck or head area, but can be located anywhere in the body. It may be discovered in a fetus during a pregnancy ultrasound, or it may be apparent at birth as a soft bulge under the skin. Herein, we present a case of neonatal mediastinal cystic hygroma extending from neck to the intrathoracic cavity causing respiratory distress that resolved spontaneously, along with a review of the literature.***

Keywords: neonatal cystic hygroma, lymphatic malformation, mediastinal mass, neonatal respiratory distress, spontaneous resolution.

1. Case Presentation

A full term female newborn product of cesarean section to a 30 year old multigravida Jordanian mother with good Apgar scores at 1 and 5 minutes, her birth weight is 2.8 kg. Antenatal ultrasound revealed a large cystic mass in the chest suggestive of congenital cystic adenomatoid malformation (CCAM). The baby was admitted to the neonatal intensive care unit (NICU) with respiratory distress on nasal CPAP (NCPAP). Physical examination showed a healthy female newborn with no apparent dysmorphic features but with a small swelling on the left side of the neck that was transilluminant, suggestive of cystic hygroma Figure 1, 2.

Chest x ray showed a homogenous cystic shadow causing compression of the right lung and extending to the left hemithorax *figure 3*. Chest CT was ordered and revealed a homogenous cystic mass extending from the neck down to the anterior mediastinum consistent with the diagnosis of cystic hygroma *figure 4, 5*. Surgical opinion was requested and the plan was to wait and see. The baby's respiratory status improved gradually until she was weaned off NCPAP then discharged home at two wks of age off oxygen and on full breast feeding. Follow up chest x ray at two months of age showed complete resolution of the mass shadow *figure 6.* Her karyotype, echocardiography, thyroid function test and abdominal ultrasound were all normal. So, far the baby is thriving very well with no complaints.

2. Discussion

Cystic hygroma (CH) is a benign malformations of the lymphatic system that can occur anywhere on the skin and mucous membranes *1*. Currently, this lesion is more commonly referred to as a lymphatic malformation (LM). It affects the head and neck in about 75% of cases with a predilection for the left side *2*. Karyotypic abnormalities are present in 25 - 70% of children with LM *3*. In our case the baby had no other abnormalities and her chromosomal study was normal. Most LMs (50 - 65%) are evident at birth, and 80 - 90% of them present by age 2 years. Cystic hygroma diagnosed after birth is usually associated with a good prognosis. The distribution of this condition is equal among males and females. Most series report no racial predominance, though a decreased incidence in African Americans has been described *4*.

The mainstay of treatment of LMs is surgical excision. Complete excision has been estimated to be possible in roughly 40% of cases. Recurrence is rare when all gross disease is removed, if any residual tissue remains, the recurrence rate is approximately 15%. Less invasive options such as sclerotherapy, oral medication and laser therapy alone or in combination can be used in small non - obstructive lesions *5*. Unlike hemangiomas, LMs do not commonly resolve spontaneously *6*.

In the neonatal population, management of the airway in cervical cystic hygromas presents with several challenges that require coordinated airway management and planning among pediatric teams. Ideally, if a critical respiratory event is anticipated prior to delivery, an ex utero intrapartum (EXIT) procedure can establish a safe, patent airway during labor without interrupting maternal - fetal circulation, thereby preventing hypoxemia and risk of hypoxic organ injury. The EXIT procedure involves delivering the head and shoulders of the fetus through a hysterotomy to maintain uteroplacental circulation; allowing time for a coordinated airway evaluation and intervention 7. Contrary to expectations, the cystic hygroma in our patient, though large and extending into the mediastinum, resolved spontaneously in a relatively short time without any adverse consequences.

This case is significant as it highlights the potential for spontaneous resolution in cases of neonatal mediastinal cystic hygroma, which could inform and potentially alter current treatment approaches.

3. Conclusion

Neonatal cystic hygromas are challenging lesions that require coordinated airway management and planning among pediatric teams. Treatment options for a cystic hygroma depend on the size, location and symptoms present. Cystic hygroma may shrink or go away without treatment as in this case. A watchful waiting for lymphatic malformations (LMs) may be considered for lesions not causing life - threatening airway compromise.

References

[1] Wassef M; Blei F; Adams D; Alomari A; Baselga E; Berenstein A; Burrows P; Frieden IJ; Garzon MC; Lopez - Gutierrez JC; Lord DJ; Mitchel S; Powell J; Prendiville J; Vikkula M; **Vascular Anomalies Classification: Recommendations From the International Society for the Study of Vascular Anomalies** Pediatrics.2015; 136 (1): e203 - 14

[2] Ueno S; Fujino A; Morikawa Y; Iwanaka T; Kinoshita Y; Ozeki M; Nosaka S; Matsuoka K; Usui N **Treatment of mediastinal lymphatic malformation in children: an analysis of a nationwide survey in Japan**. Surg Today.2018; 48 (7): 716 - 725

[3] P G Gallager, M J Mahoney, J R Gosche **Cystic hygroma in the fetus and newborn** Semin Perinatol 1999 Aug; 23 (4): 341 - 56

[4] Ersoy AO, Oztas E, Saridogan E, Ozler S, Danisman N. **An Unusual Origin of Fetal Lymphangioma Filling Right Axilla**. J Clin Diagn Res.2016 Mar; 10 (3): QD09 - 11

[5] M. J. Neidich, J. D. Prager, S. L. Clark, R. G. Elluru **Comprehensive airway management of neonatal head and neck teratomas** Otolaryngology - Head Neck Surg (Tokyo), 144 (2) (2011), pp.257 - 261

[6] Devki C. Shukla, Benjamin M. Laitman, Aldo V. Londino **Airway management in a neonatal cystic hygroma**. Otolaryngology Case Reports Volume 15, June 2020, 100157

[7] N. Zadra, F. Giusti, P. Midrio **Ex utero intrapartum surgery (EXIT): indications and anaesthetic management** Best Pract Res Clin Anaesthesiol, 18 (2) (2004), pp.259 - 271

Figure 1: Cystic mass of the Lt side of the neck

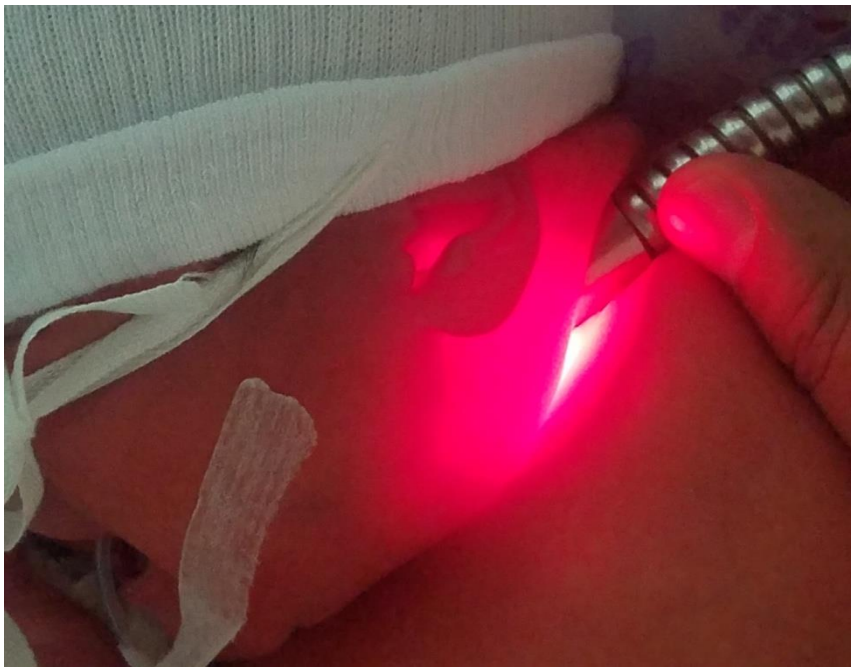

Figure 2: Transillumination positive

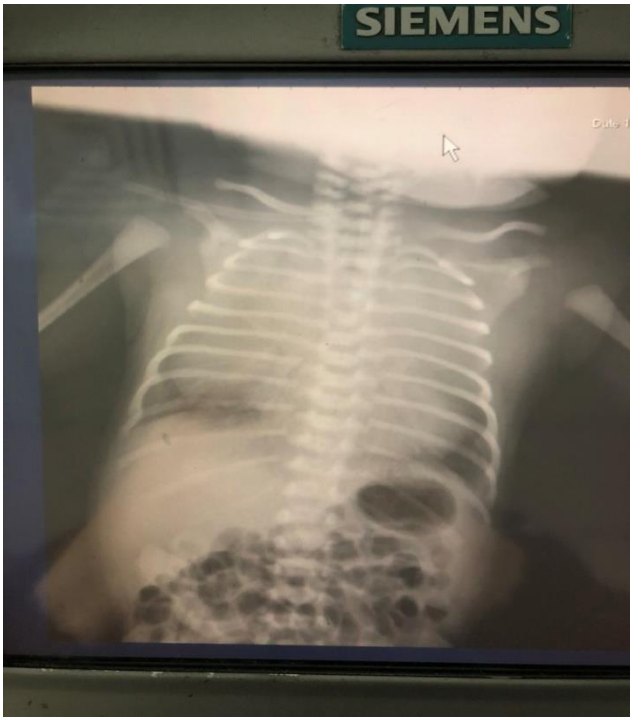

Figure 3: CXR: Bilateral dense shadows involving both lung fields

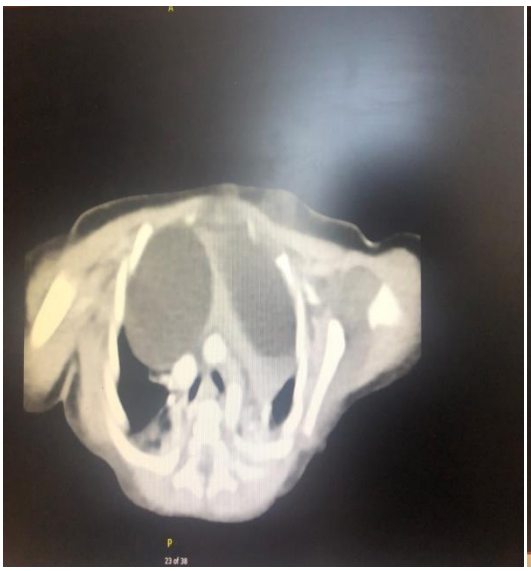

Figure 4

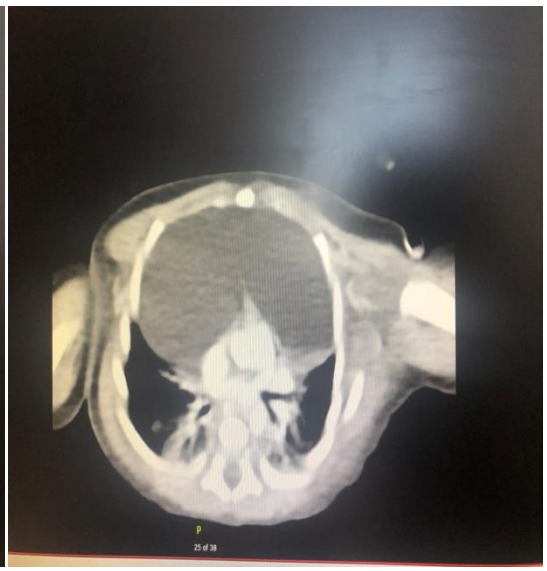

Figure 5

Chest CT: A homogeneous cystic mass involving the anterior mediastinum

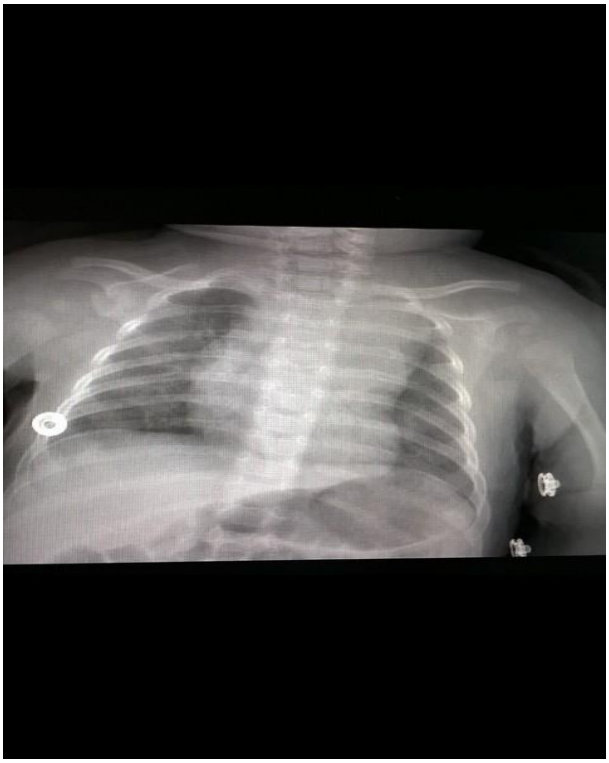

Figure 6: CXR at 2 months of age showing complete resolution of the mass shadow