ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 06 | June - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A CASE REPORT OF LYMPHOVENOUS MALFORMATION – CYSTIC HYGROMA

Pediatrics

Dr. Ajith Krishnaa Kalirajan Balakrishnan Junior Resident, (MD PEDIATRICS), Rajarajeswari Medical College And Hospital, Bangalore.

Dr Adarsh .E* M.B.B.S, DCH, MD (PEDIATRICS), Professor & Head, Department Of Pediatrics, Rajarajeswari Medical College And Hospital, Bangalore. *Corresponding Author

Dr Venkateshmurthy. D. V M.B.B.S, MD (Pediatrics), DrNB (Neonatology), Associate Professor, Department Of Neonatology, Rajarajeswari Medical College And Hospital, Bangalore.

ABSTRACT

Cystic hygromas are congenital malformations of the lymphatic drainage system that typically form in the neck which can pose an airway obstruction risk in infants. The overall incidence is 1:12000. It can be associated with other congenital anomalies such as Down's and Turner's syndromes. Early antenatal detection for safe confinement and prompt intervention following delivery plays a key role, surgical excision being the gold standard.

KEYWORDS

Cystic hygroma, congenital malformation, lymphatic system, Down's syndrome, cyst excision

OVERVIEW

Cystic hygromas are the most common subtype of lymphangiomas, presenting at birth and in early infancy. It is inherited as an autosomal recessive trait. It is potentially a life-threatening condition. The CH of the neck results from maldevelopment between the lymphatic and venous systems and manifests as mono- or multiloculated mass due to abnormal fluid accumulation inside the lymphatic vessels[1]. It predominantly occurs in the cervicofacial regions (80%), particularly at the posterior cervical triangle in infants. The condition is more common in patients with chromosomal abnormalities, such as Turner syndrome, Down syndrome[2].

Case Report

Primi mother at 39+4 weeks of gestation, booked case outside, with antenatal USG scan done at 32 weeks showed multiloculated cystic lesion 8.4 x1.7 cm over right side of fetal neck, with multiple thin and thick internal septations noted within, ?Lymphatic malformation (Fig 1 & 2) , was referred to our hospital where she delivered a Term, Male baby via LSCS (i/v/o Cystic hygroma), with birth weight 3.2 kg, baby cried immediately after birth, AGA, with APGAR 8/1', 9/5' on 25/9/23 at 10.04 am at RRMCH (Fig 3). On examination Large cystic swelling over right neck, size ~ 10 x 7cm, soft in consistency, oval shaped, fluctuatant, brilliantly transilluminant (Fig 4), with visible superficial veins. No signs of respiratory distress seen or compressing the underlying structures resulting in dysphonia or dysphagia. USG neck showed Veno-lymphatic Malformation in the right neck s/o Cystic Hygroma. Day 1 and Day 3 blood workup was normal. Metabolic workup (TMS/NBS was negative, Thyroid profile was normal). Genetic workup - Karyotyping was normal. The baby was clinically stable with no signs of airway obstruction, tolerating feeds well. To rule out associated congenital anomalies, radiological investigations Neurosonogram, Ultrasound Abdomen & Pelvis, 2D ECHO was done which showed normal study. Paediatric surgeon opinion was obtained and was planned for surgical excision of the same. As the baby was clinically stable with no signs of airway obstruction the baby was discharged on day 6 of life. The baby was doing well and was on regular follow up at high risk clinic, and has been planned for surgical excision of the cyst.

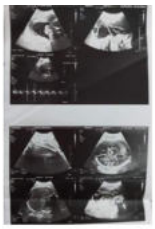

Fig 1

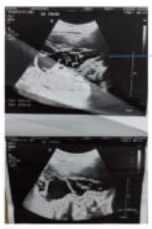

Fig 2

Fig 3

Fig 4

By 7 months of age, the infant was doing well, weight gain was appropriate gain and normal developmental milestones attained appropriate for age, the size of the lesion did not reduce and so MRI Neck (Fig 5 & 6) was done to rule out any mediastinal extension and to rule out the exact extent of the cyst which showed a well defined multiloculated cystic lesion with thin internal septations measuring 6.5 x 7.1 x 6.9 cm (TR x AP x CC) in the subcutaneous plane of lateral and posterior aspect of right cervical and supraclavicular region suggestive of Venolymphatic malformation. Few foci of T1 hyperintensity noted within the lesion-likely hemorrhagic foci, few vascular channels with flow voids in inferolateral aspect of lesion. There is minimal trans-spatial extension in the medial aspect of the lesion, underlying muscular plane appears normal.

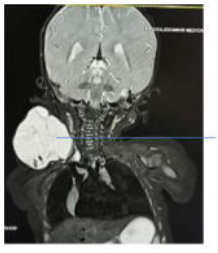

Fig 5

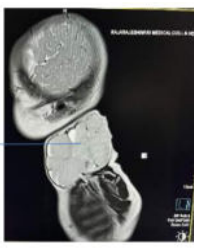

Fig 6

Pre anaesthetic workup done and pediatric fitness was obtained and the baby was taken up for surgery. Excision of the cyst and sclerosant (Bleomycin) injection into floor was done on 19/4/2024 in RRMCH. Intra op findings was multiloculated cystic lesion with hemorrhagic collection within cysts. Intra and Post operatively uneventful.

Specimen sent for Histopathological analysis (Fig 7) - Grossly grey white to grey brown soft tissue mass measuring 6 x 5 x 0.5 cm, with external surface congested with fibrofatty tissue, cut surface was

multiloculated, small cysts with septations noted, filled with hemorrhagic fluid. Microscopically showed spaces filled with proteinaceous fluid and dense lymphatic aggregates. Numerous engorged vascular spaces noted with few thick and thin veins. Stroma composed of delicate meshwork of collagen punctuated by small lymphoid aggregates and dense fibrocollagenous tissue and entrapped fibroadipose tissue which was suggestive of lymphovenous malformation. Postoperatively the child is doing well (Fig 8) with adequate weight gain and the child is on regular follow up with neonatal high risk clinic.

Fig 7

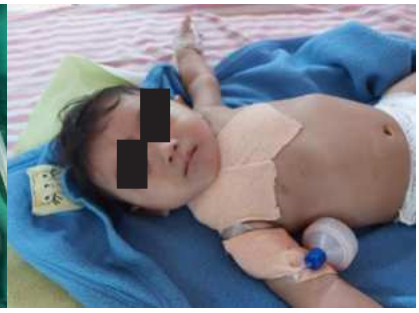

Fig 8

DISCUSSION

Cystic hygromas are congenital malformations of the lymphatic drainage system that typically form in the neck that develops at the end of the sixth week of gestation. The other sites are axilla (20%), mediastinum extension (10%), and only 1% confined to the chest. Enlargement of cystic hygromas is common, and they pose an airway obstruction risk in infants. To avoid the high mortality associated with this condition, they must be promptly diagnosed and treated[3].

It is occurs during embryonic development caused by failure of the lymhatic tissue to communicate with the central lymphatic system and venous system. Afterward, the dilatation of the isolated lymphatic tissues happen which gives the cystic morphology. It manifests as large single or multiple loculated cysts filled with serous secretions. It often appears at birth (60%), and up to 90% become overt before the age of two. It is associated with other congenital anomalies such as Downs and Turners syndromes and also heart defects[4]. The head and necks are the most common sites of origin (80%), but it also can extend into the floor of the mouth, the pharyngeal wall, and the mediastinum. The patient with cervical CH may present with dyspnea/respiratory compromise/respiratory distress due to airway obstruction or feeding problem/dysphagia.

Antenatal diagnosis of fetal cystic hygroma is essential not only for prognostication but also for planning the mode of delivery and postnatal management. Fetal echo is recommended for detecting cardiac anomalies which can bother viability[5]. Teamwork consists of obstetrician, pediatrician, pediatric surgeon, anesthesiologist, and radiologist are required.

The prenatal diagnosis using ultrasonography (USG) may shown a septated or non-septated bilateral thin-walled cystic lesion in the fetal occipitocervical region in both the sagittal and axial plane. Sometimes, CH may have mixed echo images due to the combination of cystic and solid components inside the mass. No blood flow is detected on color doppler USG. Aspiration of the lump in combination with imaging is the gold standard for diagnosing cystic hygromas[6]. Ultrasonography is the least invasive of all and typically shows multicystic lesions with internal septations. CT and MRI can be helpful in delineating the lesion further and for planning surgery as they help to illustrate the involvement and proximity to neighboring structures. Karyotyping can be done in case of any doubt of chromosomal disorder and aneuploidies. (obtained by amniocentesis, chorionic villus sampling, or umbilical blood sampling). Fetal echo is recommended for detecting cardiac anomalies which can bother viability. Chest x-ray is important for all cases to identify the intra-thoracic extension.

Treatment options will be individualized depending on the size, anatomical location, and complications of the lesion. The mainstay of management is surgical[7]; however, other options include Intralesional sclerotherapy (OK-432, Rapamycin (Sirolimus), drainage, radio-frequency ablation, or cauterization. Cystic hygromas that develop into abscesses will require antibiotics, antipyretics, and analgesia, with or without subsequent surgical management.

REFERENCES

1. L. Schreurs, L. Lannoo, L. de Catte, D. van Schoubroeck, K. Devriendt, J. Richter First trimester cystic hygroma colli: retrospective analysis in a tertiary center Eur J Obstet Gynecol Reprod Biol, 231 (2018), pp. 60-64, 10.1016/j.ejogrb.2018.10.019.
2. B. Mirza, L. Ijaz, M. Saleem, M. Sharif, A. Sheikh Cystic hygroma: an overview J Cutan Aesthetic Surg, 3 (2010), p. 139, 10.4103/0974-2077.74488.
3. K. Rao, T. Shenoy Anesthetic management of a large cystic hygroma in a newborn Anesth Essays Res, 9 (2015), p. 270, 10.4103/0259-1162.156364.
4. Damaskos C, Garmpis N, Manousi M, Garmpi A, Margonis GA, Spartalis E, Doula C, Michail-Strantzia C, Patelis N, Schizas D, Arkoumanis PT, Andreatos N, Tsourouflis G, Zavras N, Markatos K, Kontzoglou K, Antoniou EA. Cystic hygroma of the neck: single center experience and literature review. Eur Rev Med Pharmacol Sci. 2017 Nov;21(21):4918-4923.
5. Bloom DC, Perkins JA, Manning SC. Management of lymphatic malformations. Curr Opin Otolaryngol Head Neck Surg. 2004 Dec;12(6):500-4.
6. Gallagher PG, Mahoney MJ, Gosche JR. Cystic hygroma in the fetus and newborn. Semin Perinatol. 1999 Aug;23(4):341-56.
7. Ha J, Yu YC, Lannigan F. A review of the management of lymphangiomas. Curr Pediatr Rev. 2014;10(3):238-48.