

UNIT OF AXIS JOURNALS

International Peer Reviewed Medical Journal Committed for Excellence

Cystic Hygroma in a Newborn

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CASE REPORT

ABSTRACT

A newborn presented with cystic hygroma in axilla. In view of rarity of condition, presentation at birth at unusual site and classical features of clinical condition, we are reporting the case.

Key Words

Axillary swelling, cystic hygroma, newborn

Received on 18 April 2014

Accepted on 11 May 2014

Published on 22 May 2014

INTRODUCTION

Cystic hygroma is a benign congenital lymphatic malformation that has its origin in the hypoplastic communicating channels between lymphatic systems and their draining veins. The incidence of cystic hygroma is approximately 1/6000 live births¹. 70–80% of cystic hygromas occur in the neck, usually in the posterior cervical triangle². They are soft, cystic, compressible, of variable size but characteristically brilliantly transilluminant. Cystic hygroma is known to present at birth in about 50% of the affected newborns and 90% present by age 2 years³. their course is indolent in most cases^{4,5}. However, these lesions may hemorrhage, develop inflammation or infection, or may progressively enlarge, leading to an expanding lesion that may physically compress local organs⁶. Surgical excision is regarded as the treatment of choice.

We report here a case of cystic hygroma with the classical presentation but at an uncommon site.

CASE REPORT

A one day old 2.7kg, term, male newborn was admitted with complaints of a swelling in the right axillary region. The infant was delivered at a community hospital and his general condition was good. A 5.0 x 4.0 cm swelling extending from the axilla to mid-thoracic region. (Figure1)



Figure 1; Newborn with cystic hygroma in right axilla.

It was soft in consistency, fluctuant, non reducible, with smooth surface, rounded margins, normal overlying skin, freely movable, cystic and transillumination test was positive. (Figure 2)



Figure 2; Classical transilluminant cystic hygroma in right axilla of newborn.

The temperature over swelling was normal and it was not indurated. Ultrasonography (USG) of the swelling showed multiseptate cystic lesion in the subcutaneous plane with no internal vasculature. The case was consulted with pediatric surgeon and ideal time of excision was planned. The baby was discharged after 5 days on exclusive breast feeding and called for follow up at 6 months of age.

DISCUSSION

This case is discussed here because of classical swelling, at less common site presenting at birth. Cystic hygromas or lymphangiomas are believed to occur as a result of the failure of establishment of appropriate connection to the normally present lymphatic channels. Lymphangiomas may be divided histologically into two major groups based on the depth and the size of abnormal lymph vessels. The superficial ones are called lymphangioma circumscriptum. The more deep seated ones are cavernous lymphangioma or cystic hygroma⁷. Cystic hygroma was first described by Wernher in 1843.^{8,9} The most common site is the head and neck (75%) with more predilections over the left side.⁸ With in the neck, the posterior triangle is most commonly affected. This predication is related to embryological development of lymphatic tracts. In 20% cases, it may occur in axilla, and rare sub sites are superior mediastinum, chest wall, mesentery, retro-peritoneal region, pelvis and lower limbs¹⁰. It is described as a congenital multiloculated lymphatic lesion containing watery fluid. They are soft, vary in size and shape, and tend to grow extensively if not surgically excised¹¹. They are most commonly found at birth although some may occur after birth.⁸ Cystic hygroma is known to present at birth in about 50% of the affected newborns and 90% present by age 2 years³. It can be found in normal babies as well as in babies born with chromosomal anomalies like Turner syndrome, Noonan syndrome, trisomies, fetal alcohol syndrome, chromosomal aneuploidy, cardiac anomalies and fetal hydrops¹². Although cystic hygromas tend to enlarge progressively over months a relatively rapid increase in size has also been described¹³.

Cystic hygromas need to be differentiated from deep seated hemangiomas, soft tissue tumours like teratoma (very rare), lipoma. Cystic hygroma of the axillary space must also be differentiated from high thoracic myelomeningocele¹⁴. The latter usually has a cystic structure containing only cerebrospinal fluid and neural elements (myelomeningocele). The diagnosis of lymphangiomas is usually made solely on the basis of history and physical examination. USG with Doppler flow is cost-effective; but operator dependent and does not accurately depict relation of the lesion with surrounding structures. CT Scan/ MRI is used to confirm the diagnosis and to look for intrathoracic component. Biopsy is performed if malignancy is suspected. The management of lymphangiomas including cystic hygromas is preferably surgical, although a careful "wait and see" policy may be indicated in few asymptomatic cases, as spontaneous regression has been reported for as long as 18-24 months¹⁵. However immediate surgery is indicated with signs of infection and for lymphangiomas located over tongue, vocal cord, eyes, ears, nose interfering with the functioning of the organ. Other treatment modalities include aspiration, radiation, and injection of sclerosing agents, in particular the agent OK-432, derived from a strain of streptococcus pyogenes, which has been used successfully, especially in macrocystic lymphangiomas and in patients who are at increased anesthetic risk.¹⁶

CONCLUSION

Cystic hygroma should always be considered in the differential diagnosis of cystic lesions with onset at birth. This case highlights the importance of clinical history and examination in diagnosing this rare entity. It emphasizes that conventional physical signs (like transillumination) peculiar for it, can substitute unaffordable and unavailable sophisticated tools in countries with limited resources.

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Please cite this paper as: Aggrawal R, Naik T, Ambey R, Gaur A. Cystic Hygroma in a Newborn. *Inter J Medical Sci Res Prac* 2014; 1 (2): 20-22.

ACKNOWLEDGEMENTS

Nil

PEER REVIEW

Double Blinded externally peer reviewed.

CONFLICTS OF INTEREST

Nil

FUNDING

Nil