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Case Report

Axillary Cystic Hygroma in a Three Month old infant

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Introduction

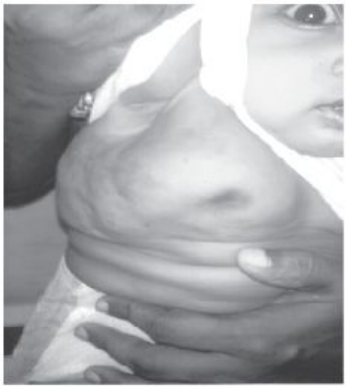
Cystic hygroma (CH) is a congenital lymphatic malformation occurring in different parts of body, typically in the region of the neck, axilla, abdominal wall, mediastinal, inguinal and retroperitoneal areas.[1,2] Cystic hygroma may result from a complete or incomplete obstruction in the lymphatics that prevents communication with the venous system and causes cysts.[3] About 75% of the cystic hygroma occur in the neck, With a predilection for the left side, mainly in the posterior triangle, 10 -20% are located in the axilla and less than 10% are located in the extremities, trunk, abdomen, genitalia, etc.[4,5] Occasionally cystic hygroma is inherited as an autosomal recessive disorder. Most common cause is idiopathic. Some of the other causes are maternal viral infection such as parvovirus and maternal substance abuse like alcohol.[6]

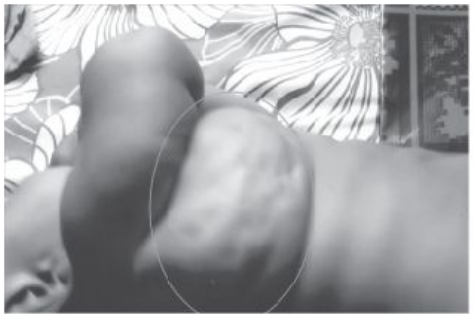
This case report is to present the foetal axillary cystic hygroma diagnosed just after delivery in a 30-yr-old woman.

Case Report

A 3 month old male baby, first issue of a non-consanguineous parents coming from a middle class family of Dhaka got admitted to hospital with the complaints of fever for last 5 days, and a mass in the axillary region of right side since birth. The mother was under regular antenatal checkup. Her antenatal period was uneventful. The baby was delivered at term in a hospital by lower uterine cesarean section (L.U.C.S).

The baby was born with a mass in the axillary region but had no other problems. He was on exclusive breast feeding and was immunized as per EPI schedule. On clinical examination, a large multi-loculated soft cystic, painless mass in the right axilla that measured 12 x 15 cm sized, and extended to the right anterior chest wall was found. Parents noticed that initially the mass was small but gradually increased to attain the present size. No other anomalies were noted. Ultrasound examination performed in our hospital, revealed multi-septated cystic mass in right axilla that measured 10 x 14 cm and extended to the right chest wall but no internal extension of lesion was seen at present which was suggestive of cystic hygroma. Patient was normal karyotype (46, xy). The chest radiograph did not show any abnormality. Since this patient was febrile, he was diagnosed as infected cystic hygroma and was treated conservatively with antibiotics, and antipyretics. Aspiration of cystic hygroma was done, both for diagnosis and treatment at about 3 months of age. Also Sclerotherapy with intra-lesional Bleomycin has been tried at 4 month of age. Since this patient did not improve with these management, it was decided to go for surgical excision.





Discussion

Hygroma in Greek means water containing Tumour. Cystic hygroma is a benign congenital malformation of the lymphatic system that has its genesis in the lack of development of communication between the lymphatic and venous systems. 80% of cystic hygromas occur in the neck, 10-20% are located in the axilla, and less than 10%

are located in the extremities, trunk, abdomen, genitalia, etc.[7] Cystic hygroma may result from a complete or incomplete obstruction in the lymphatic that prevents communication with the venous system and causes cysts. The hygroma describes an endothelial lined mass consisting of small to medium sized lumina containing lymphatic fluid, together with a mixture of loose collagen tissue, adipose tissue and occasionally, vascular tissue. The cysts may be unilocular but more often the structure contains multiple cysts infiltrating the surrounding structures and distorting the local anatomy. The septated cystic hygroma may result from a complete obstruction in the lymphatic sacs that prevents communication with the venous system and causes lymph fluid to accumulate and dissect into tissues, thus creating large multilocular cysts. Nonseptated cystic hygroma may result from a temporary accumulation of lymph fluid due to incomplete obstructions of lymphatic drainage. Axillary cystic hygroma was rarely reported and detected often in mid-gestation period. Foetal cystic hygroma has been associated with foetal aneuploidy, hydrops foetalis, structural malformations and intrauterine foetal death. It is reported to occur between 1 in 6000 and 1 in 16,000 live births but it is estimated to be much more than this proportion as we take into account the abortions.[8]

Two distinct categories of foetal cystic hygroma have been described: Those diagnosed in later pregnancy, which tend to be isolated lymphangiomas, and those diagnosed in early pregnancy, which are commonly associated with other malformations. Retrospective case series describing the prenatal diagnosis of this condition suggest that those diagnosed early in pregnancy are associated with a poor prognosis. The cystic hygroma in later gestation, which likely represents a lymphangioma that is not associated with either aneuploidy or other foetal malformations, and can be expected to yeild good prognosis. Cystic hygroma incidence is equal in both sexes.[9] The frequency of a chromosomal abnormality associated with cystic hygroma may be as high as 78% Turner syndrome being the most common. Prognosis for cystic hygroma is grim if the Karyotype is abnormal.[10] The usual presentation of cystic hygroma apparent at birth is a painless mass with worries and queries of the parents about the lesion. The other modes of presentations are related to the complication or effects of cystic hygroma, such as respiratory distress, feeding difficulty, fever, sudden increase in the size and infection in the lesion. On clinical examinations, these lesion appear soft, compressible, non tender, transluminant and without any bruit.[11]

Foetal axillary cystic hygromas have been reported rarely and usually as a sonographic finding in mid-gestation. Axillo-thoracic cystic hygroma may be diagnosed during routine antenatal ultrasound follow-up. On an ultrasonological scan, it appears as a hypo-echogenic multilocular cystic mass with septa of variable thickness. MRI (magnetic resonance imaging) is important in determining the characteristics of the tissue and tumour extent. Considering its association with several chromosomal anomalies, determination of foetal Karyotype may be undertaken for providing accurate diagnosis and genetic counseling. Repeat sonological evaluation may be necessary antenatally for the evaluation of the tumour growth. As cystic hygromas are known to lead to obstructed labour, and neonatal asphyxia, an elective caesarean section should be considered as the preferred mode of delivery.[7] Differential diagnosis of Axillothoracic cystic hygroma, simple cyst, hemangiomas, branchial cleft cysts, hemongio-lymphangiomas, lymphocele, teratomas, ectopia cortis, Klippel-Trauray syndrome should be considered.[8] In this case report ante-natal ultrasonography was done after mid-gestation but foetal axillary cystic hygroma could not be detected and it was diagnosed after birth.

The most preferred modality of treating cystic hygroma remains complete surgical excision; however, many recent case reports have increasingly documented remarkable results for management of such lesions with sclerosant agents. Sclerotherapy with intra-lesional bleomycin as a primary treatment modality, for cystic hygroma, has been tried. Various case reports and original studies have documented good response to the therapy. The other agent used as sclerosant is OK 432, has more satisfactory results and less complication as compared to bleomycin. The other treatment modalities that have been employed with variable results include simple drainage, aspirations, radiation, laser excision, radio-frequency ablation and cauterization, which are contraoversial.[11]

Conclusion

The incidence of diagnosis of foetal cystic hygroma has increased due to routine antenatal ultrasound screening. Antenatal diagnosis of foetal cystic hygroma helps in planning a better anticipatory care (elective caesarean section, conducting delivery in a well-equipped center and undertaking investigations for the diagnosis of other associated congenital anomalies and chromosomal abnormalities). Cystic hygroma is a manageable lesion in the paediatric population. Suitable treatment should be

opted, based on case to case variation. Preoperative imaging for diagnosis and searching for intrathoracic extension is essential. Optimum treatment can be given either by surgery or sclerotherapy or combined use of both. Recently invented treatment modalities such as laser and radiofrequency can also be used in selected patients. Surgical excision is the treatment of choice.

Reference

1 . Pijpers L, Reuss A, Stewart PA, et al. Fetal Cystic Hygroma: Prenatal Diagnosis and Management. Obstet Gynecol.1988; 72: 223-24 [Pub Med].

2 . Cohen MM, Schwartzs, Schwartz MF, et al. Antenatal detection of Cystic hygroma. Obstet Gynecol Surv. 1989; 44: 481 – 90. [Pub Med].

3 . Brumweld CG , Wenstrom KD , Davis Ro , et al . Second Trimester Cystic hygroma: Prognosis of septated and non-septated lesions. Obster Gynecol. 1996; 88: 979 – 82. [Pub Med].

4 . Mc Coy Mc, Kuller JA , chescheir NC, Coulson CC , Katz VL , Naka- Yama DK. Prenatal diagnosis and management of Massive bilateral axillary Cystic Lymphangioma. Obstet Gynecol 1995; 85: 853-6.

5 . Judith E,Allanson MD . Lymphangioma. In: Stevenson RE, Hall JG, Goodman RM, Editors, Human Malformation and Related Anomalies. New York: Oxford University Press, 1993; 288.

6 . Gnanavel A, Divya P and Vikram T. Cystic Hygroma –A case Report and its Embryological basis. Anatphysiol 2015; 5:1.

7 . Manikoth P, Mangalore GP, Megha V.Axillary Cystic Hygroma. J postgrad Med 2004; 50: 215-6.

8 . Temizkan O, et al. Fetal Axillary Cystic Hygroma: A Case Report and Review. Rare Tumors 2011 Oct. 21; 3 (4): 39.

9 . Langer JC ,Fitzgerald PG , Desa D ,et al. Cervical Cystic Hygroma in the Fetus: Clincal Spectrum and outcome .J Pediatr Surg .1990; 25: 58-62 [pub Med].

10. Carr R F, Ochs RH ,Ritter DA, Kenny JD, fridey JL, Ming P M .Fetal Cystic Hygroma and Tunner's Syndrom. Am J Dis child 1986; 140: 580-3.

11. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Cystic Hygroma: An Overview. J Cutan Aesthet Surg. 2010; 3 (3):139-144.