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Case Report: Fetal cystic hygroma- Does it have a normal outcome?

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Abstract

Cystic hygroma is a benign congenital malformation of the lymphatic system. Fetal cystic hygromas are usually associated with chromosomal anomalies and mostly have an adverse outcome. The mean size of cystic hygroma is 8 cm. We report a case of a giant cystic hygroma (17.1 ×11.3×8.1 cm) in a newborn which was diagnosed antenatally and had no syndromic association. It was successfully operated and baby remains free of any recurrence or complications at 2 months of age.

Introduction

Cystic hygroma, also known as cystic lymphangioma, is a benign congenital malformation of the lymphatic system. Incidence of Cystic hygroma is approximately 1/6000 live births.(1) Although these lesions can involve any part of the body, 70-80% are located in the cervico-facial region; commonest location being the posterior cervical triangle.(2) About 78% of such cases are associated with chromosomal anomalies, the commonest being Turner's syndrome (58%). The remaining cases with normal chromosomal analysis are associated with various physical anomalies.(3,4) 50% of these lesions present at birth and around 90% manifest by 2 years of age.(5) Cystic hygroma vary from 1-30 cm in size, the mean size in Stromberg's series was 8 cm.(6) Fetal Cystic hygromas are rare and they often progress to hydrops and cause fetal death. The survival rate of antenatally diagnosed Cystic hygromas has been reported as 2-3%.(4,7) We report a case of antenatally diagnosed large Cystic hygroma (17.1×11.3×8.1 cm) in a newborn female, which required surgical excision at birth, and had a normal outcome till date. Chromosomal analysis ruled out any syndromic association. The baby did not develop any neural paresis or paralysis, which are common complications post surgical excision.

Case Report

A late preterm (36+4 weeks) was delivered by LSCS to a primi mother at a tertiary care institution. Fetus was antenatally diagnosed with a large cystic swelling involving the left side of the neck on antenatal ultrasound. The birth weight of the baby was 3.2 kg (with cystic mass). Baby cried immediately after birth, Apgar being 8 and 9 at 1min and 5 minutes of life, respectively. Baby was admitted in NICU. MRI neck was done which was suggestive of a large lobulated cystic lymphangioma, measuring 17.1(CC) by 11.3(T) by 8.1(AP) cm (Fig. 1), arising from the posterior cervical space. There was no evidence of deep visceral space involvement, major airway/ vascular compression or mediastinal extension. Pediatric surgery consultation was taken and baby was operated on day 3 of life, due to its large size (17.1×11.3×8.1). Complete surgical excision was done and neck drain was left in situ. Baby was initially started on orogastric feeds and by day 6 of life spoon feeds were tried and baby accepted them well. Postoperative course remained uneventful. Neck drain was removed on day 10 of life. Karyotype sample sent for Chromosomal analysis was reported as normal; thus the possibility of Turner's syndrome was ruled out. Baby was discharged on breast feeds at day 12 of life. Neurological examination was normal with no neural paresis/ paralysis. Currently baby is 2 months of age with no recurrence or any neurological complication.

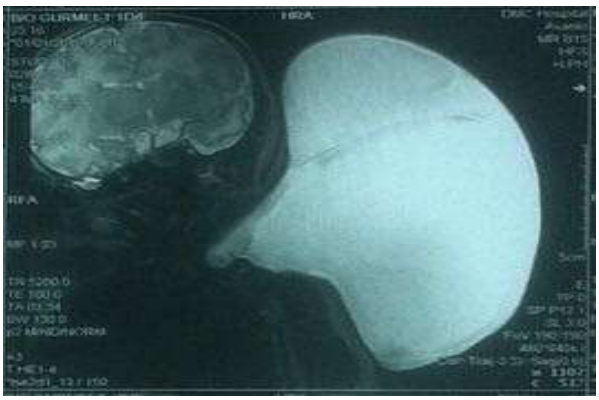

Fig. 1: MRI depicting the lesion

Fig. 2: Baby at the time of birth

Fig. 3: Baby at the time of discharge

Discussion

We report a case of a fetal cervical cystic hygroma, which despite being large, was totally asymptomatic and had a very favorable outcome. Cystic hygromas are known to arise from sequestration of lymphatic tissue from lymphatic sacs.[8] The inability to establish a communication with the remainder of the lymphatic or venous system, leads to the dilatation of these sequestered tissues and hence the cystic lesions.[8,9] These lesions can be congenital or acquired and etiology is usually multifactorial. Clinically they appear as soft, compressible, brilliantly Trans illuminant, and non-tender lesions without any bruit. Very few hygromas extend into the mediastinum. Based on the size of the cysts contained, they are classified as micro cystic (<2cm), macro cystic(>2cm) and mixed.[10] They can also progressively increase in size causing compression of surrounding structures leading to severe respiratory and neurovascular compromise.[11,12] They tend to grow extensively if not surgically excised and are notorious for re-occurring post excision. CT scan, Ultrasound and MRI are the diagnostic modalities, which help in ascertaining the extent of the lesion and involvement of surrounding structures. In Antenatal scans, these lesions appear as multicystic lesions with or without internal septae. No blood flow is evident on performing color Doppler.[13,14] These multicystic homogenous lesions may be filled with milky, serous or straw colored fluid.[15] The fetus with cystic hygromas can be associated with anomalies like Turner's syndrome, Down's syndrome, Trisomy 18, Trisomy 13, Noonan syndrome, cardiac anomalies and fetal hydrops.[16-18] The complications associated with Cystic hygroma are secondary infection, spontaneous hemorrhage, airway compromise and dysphagia. Although cosmetic deformity is usually the reason of treatment, small and asymptomatic lesions can be monitored over time. Complete surgical excision remains the chief modality of treatment; extension into the mediastinum and involvement of surrounding neurovascular structures can pose a difficulty in their identification and excision. Post excision neural paralysis or paresis can occur; infection, hemorrhage and discharge being the other common complications. There is a 20% chance of recurrence following complete excision; hence a combined approach of treatment is preferred.[19,20] Various studies have documented good response to sclerosants like Bleomycin[21,22] and the recent OK432; the latter having a better safety profile but remains experimental.[23] The other modalities of treatment are aspiration using a wide bore needle, radiotherapy and sclerotherapy.[24-27] A multidisciplinary approach will help obtain a successful outcome.

Conclusion

This is a rare case of a large cystic hygroma which was antenatally diagnosed and had a good outcome. There was no syndromic association and no physical anomalies were present. The baby did not develop any complication post-surgical excision and remains asymptomatic till 2 months of age.

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