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CASE REPORT

Fetal Axillary Cystic Lymphangioma Detected by Prenatal Ultrasonography

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Fetal cystic lymphangioma, commonly called hygroma, is formed by multiple cysts ranging from few millimeters to several centimeters in size. Fetal cystic lymphangioma is a rare developmental congenital anomaly of the lymphatic system. Most are found in the neck and axilla, rarely in the retroperitoneum, abdominal viscera, limbs, bones, and cervico-mediastinal area. The prenatal diagnosis of cystic hygroma by ultrasound guidance is commonly localized in the nuchal region. We report a case of rare fetal axillary hygroma that was diagnosed at 27 weeks of gestation and delivered at 37 weeks of gestation.

Keywords: Cystic lymphangioma; Axillary; Fetal

INTRODUCTION

Hygroma in Greek means water-containing tumor. Fetal cystic hygromas are congenital malformations of the lymphatic system appearing as single or multiloculated fluid filled cavities [1]. About 75% of the tumors occur in the posterior triangle of the left neck and 20% of the tumors occur in the axillary region [2-4]. About 50% of these appear at birth, and 90% appear until age two. It is mainly associated with Turner's syndrome and is accompanied by various structural deformities [5,6]. Our patient was a normal female karyotype, and there were no structural abnormalities other than axillary cystic hygroma.

Recently, we experienced a rare case of fetal axillary cystic hygroma at 37 weeks' gestation in a 36-year-old multiparous woman. Here we report this case with a brief review of the literature.

CASE REPORT

A 36-year-old woman with gravida 3 and para 2 was referred to Soonchunhyang University Cheonan Hospital at 27 weeks' gestation because of a mass on the left fetal chest wall detected by routine ultrasound guidance (USG) performed at a private clinic at 27 weeks' gestation. There was no fetal abnormality by ultrasonography at 25 weeks' gestation at a private clinic. The patient's family history and previous medical history were unremarkable. Her first normal child was delivered by vaginally 5 years ago and second normal child was delivered vaginally 2 years ago. In the current pregnancy, the maternal serum alpha fetoprotein checked at 16+5 weeks' gestation was normal. USG examination, performed in our hospital at 27 weeks' gestation, revealed a 6×5 cm-sized, multiseptated cystic mass in the left axillary region of the fetus (Fig. 1). Amniotic fluid amount was normal and no other structural abnormalities were found with normal echocardiography in the fetus. The biparietal diameter, head circumference, femur length, and abdominal circumference in the fetus corresponded to 27 weeks' gestation. With the diagnosis of a fetal axillary cystic lymphangioma, we performed follow-up examinations every 2 weeks with USG. The size of the fetal axillary mass was slightly increased with increasing size of the fetus. However, there were no findings such as fetal hydrops. She was delivered vaginally at 37 weeks of pregnancy after regular labor followed by premature rupture of membranes. A 2,720 g-sized female infant was delivered with Apgar scores of 9 and 10 at 1 minute and 5 minutes, respectively. The fifth day after delivery, the baby demonstrated a 6×7 cm-sized, soft cystic mass in the left axillary area (Fig. 2). No other structural anomalies were identified. Umbilical cord blood

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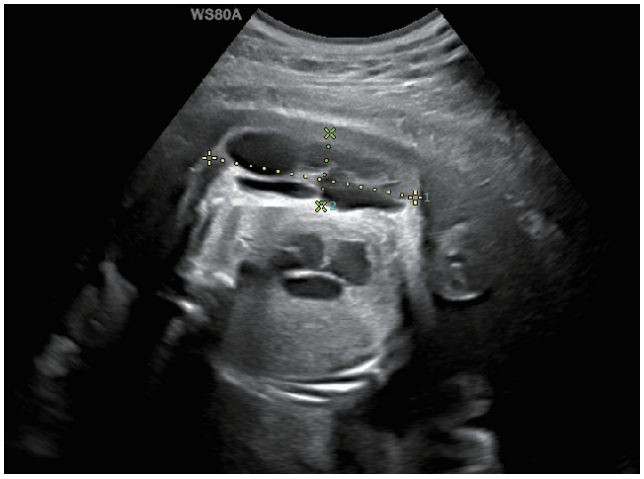

Fig. 1. Transverse sonographic view of chest at 27 weeks' gestation demonstrating a large cystic mass with septum in the left axillary region.

Fig. 2. The fifth day after delivery, the baby demonstrated a 6 × 7 cm-sized, soft cystic mass in the left axillary area.

taken at the delivery revealed a normal karyotype (46, XX). Chest USG was performed on the second postnatal day (Fig. 3). It showed an 8 × 6 cm-sized, well-marginated cystic mass with multiple septation at the left lateral chest wall without extension to the upper neck or mediastinum. Operation was performed on the 6 months after birth at the department of plastic surgery and general surgery and the mass was excised. The baby showed normal recovery after surgery without postoperative complications. Currently, the baby grows well at the age of 1 year with 10.5 kg of body weight. There have been no signs of complications.

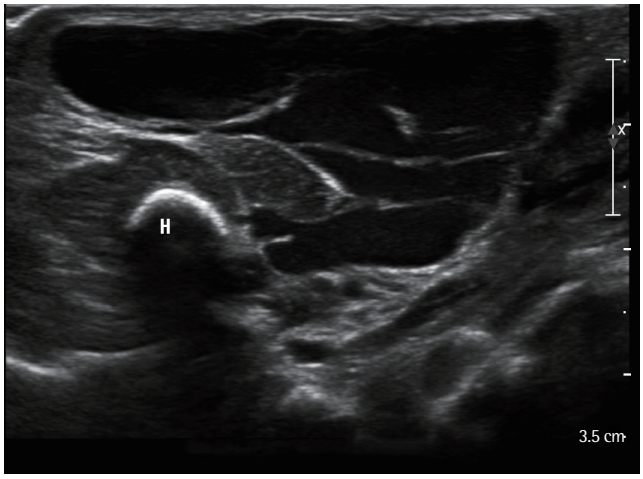

Fig. 3. Chest ultrasound demonstrating 8 × 6 cm-sized, well-marginated cystic mass with multiple septation at the left lateral chest wall without extension to the upper neck or mediastinum. H, humerus.

DISCUSSION

The incidence of cystic lymphangioma is 1:6,000 pregnancies. It is relatively common in aborted fetuses with an incidence of 1: 875 [1]. There are many case reports of cystic hygroma in the neck, but there are only a few papers about fetal axillary cystic hygroma [2,3] and few domestic papers [4]. There is no difference by sex [5,6]. Cystic hygromas are generally bilateral, thin walled, unilocular or multilocular cysts with clear to turbid fluid. Clinically the term cystic hygroma is used to refer to subcutaneous cystic spaces in the neck or to loculated cystic lesions in the noncervical regions, which have a similar appearance to the nuchal cysts. Several reviews on cystic hygroma have documented the postnatal anatomic variability [7-10]. Seventy-five to ninety percent are found in the neck, 10%–20% are located in the axilla, and less than 10% are located in the extremities, trunk, abdomen, and genitalia. Fetal neck cystic hygromas are thought to be etiologically distinct from other cystic lymphangiomas and are believed to result from inadequate drainage of the lymphatic vessels into the venous system secondary to atresia. In contradistinction, cystic lymphangioma at other locations probably develop as the lymphatic anlage grow abnormally and never achieve sufficient anastomoses with the larger lymphatic channels [11,12]. Association with other anomalies and

extension to adjacent structures leading to hydrops fetalis is frequent findings. Fortunately, this fetus did not have other deformities and hydrops. The frequency of a chromosomal abnormality associated with cystic hygroma may be as high as 78%, Turner syndrome being the most common [13]. Prognosis for cystic hygroma is grim if the karyotype is abnormal, or if ascites and pleural fluid are present, or if bilateral pleural effusions are seen. Our case showed a normal karyotype.

The survival rate progressively improves with normal karyotype, unilateral pleural effusions, atypical location, and resolution of cystic hygroma [14]. Prenatal serial USG examinations are mandatory to evaluate any changes in the clinical manifestation. Once a cystic hygroma is detected, a careful search is indicated for fetal skin edema, ascites, pleural and pericardial effusions, and cardiac or renal anomalies. Meticulous surgical excision is the treatment of choice when lesions are large. The operation is essentially conservative, since there is no justification for sacrificing any vital structures to achieve complete removal of the benign lesion. Most surgeons agree that the cystic hygroma should be excised. Conservative management with observation has been recommended by some surgeons in asymptomatic patients [15]. Because only small numbers of fetal cystic hygroma have been reported so far, it is uncertain if axillary cystic hygromas carry a marked risk for aneuploidy. If important organs are involved in neonatal surgery, it is recommended that the baby be operated 6 months or 1 year after the baby is fully grown [15]. Other treatments include radiation therapy, injection of sclerosing agents, aspiration, and carbon dioxide laser but there are still many controversial opinions about the effect [16]. Six months after birth, the patient is treated surgically and is healthy without complications after removal. Recently, we experienced a rare case of fetal axillary cystic hygroma at 37 weeks' gestation in a 36-year-old multiparous woman. Here we report this case with a brief review of the literature.

REFERENCES

1. Manikoth P, Mangalore GP, Megha V. Axillary cystic hygroma. J Postgrad Med 2004;50:215-6.
2. Kocher HM, Vijaykumar T, Koti RS, Bapat RD. Lymphangioma of the chest wall. J Postgrad Med 1995;41:89-90.
3. McCoy MC, Kuller JA, Chescheir NC, Coulson CC, Katz VL, Nakayama DK. Prenatal diagnosis and management of massive bilateral axillary cystic lymphangioma. Obstet Gynecol 1995;85(5 Pt 2):853-6.
4. Song TB, Kim CH, Kim SM, Kim YH, Byun JS, Kim EK. Fetal axillary cystic hygroma detected by prenatal ultrasonography: a case report. J Korean Med Sci 2002;17:400-2.
5. Hoffman-Tretin J, Koenigsberg M, Ziprkowski M. Antenatal demonstration of axillary cystic hygroma. J Ultrasound Med 1988;7:233-5.
6. Ninh TN, Ninh TX. Cystic hygroma in children: a report of 126 cases. J Pediatr Surg 1974;9:191-5.
7. Sheth S, Nussbaum AR, Hutchins GM, Sanders RC. Cystic hygromas in children: sonographic-pathologic correlation. Radiology 1987;162:821-4.
8. Judith E, Allanson MD. Lymphangioma. In: Stevenson RE, Hall JG, Goodman RM, editors. Human malformation and related anomalies. New York (NY): Oxford University Press; 1993. p. 288.
9. O'Brien WF, Cefalo RC, Bair DG. Ultrasonographic diagnosis of fetal cystic hygroma. Am J Obstet Gynecol 1980;138:464-6.
10. Cohen MM, Schwartz S, Schwartz MF, Blitzer MG, Raffel LJ, Mullins-Keene CL, et al. Antenatal detection of cystic hygroma. Obstet Gynecol Surv 1989;44:481-90.
11. Siegel MJ, McAlister WH, Askin FN. Lymphangiomas in children: report of 121 cases. J Can Assoc Radiol 1979;30:99-102.
12. Bhattacharyya NC, Yadav K, Mitra SK, Pathak IC. Lymphangiomas in children. Aust N Z J Surg 1981;51:296-300.
13. Carr RF, Ochs RH, Ritter DA, Kenny JD, Fridey JL, Ming PM. Fetal cystic hygroma and Turner's syndrome. Am J Dis Child 1986;140:580-3.
14. Anderson NG, Kennedy JC. Prognosis in fetal cystic hygroma. Aust N Z J Obstet Gynaecol 1992;32:36-9.
15. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Cystic hygroma: an overview. J Cutan Aesthet Surg 2010;3:139-44.
16. Bozkaya S, Ugar D, Karaca I, Ceylan A, Uslu S, Baris E, et al. The treatment of lymphangioma in the buccal mucosa by radiofrequency ablation: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2006;102:e28-31.