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Prenatal Sonographic Diagnosis of Cystic Hygroma Colli

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ABSTRACT

Objective To evaluate the role of prenatal sonography in identifying characteristics of fetal cystic hygroma.

Design Case series.

Setting Department of Obstetrics and Gynaecology, Faculty of Medicine, Maharaj Nakorn Chiang Mai Hospital, Chiang Mai University.

Subjects A total of 12 fetuses with prenatal diagnosis of cystic hygroma were evaluated and followed.

Results All cases were correctly diagnosed sonographically during the first half of pregnancy. All showed asymmetric, multiseptate cystic masses (mostly thin-walled) at posterolateral aspects of the neck. Midline septation representing the nuchal ligament extending from the posterior neck outlined by bilateral cysts were identified in all cases and may be the most specific sign for diagnosis of cystic hygroma. Fifty-eight percent (7 cases) had already developed some degree of hydrops fetalis. Fifty-eight percent (7 cases) had oligohydramnios and only one case had polyhydramnios. Chromosome studies were done in 7 fetuses, 3 had chromosomal abnormalities, 45 X0 (2 cases) and trisomy 21 (1 case).

Conclusion This small series indicates that ultrasound had high predictive value in diagnosis of cystic hygroma. The most associated abnormalities were hydrops fetalis and oligohydramnios.

Key words : cystic hygroma, prenatal diagnosis, ultrasound

Cystic hygroma is a congenital malformation of the lymphatic system. It is the most common neck mass identified in the fetus. The incidence of cystic hygroma is not well documented. Reports ranged from 1 in 6,000 pregnancies to 1 in 120 pregnancies at risk for having this structural anomaly.[1-3]

The fetal lymphatic vessels drain into two large sacs lateral to the jugular veins. If the lymphatic and venous structures fail to connect, the jugular lymph sacs will be enlarged, resulting in the dilatation of the sacs into cystic hygromas of the posterior triangles of the neck which may lead to jugular lymphatic obstruction and hydrops fetalis.[4]

The cysts are characteristically found at the posterolateral region of the neck and are frequently divided by random, incomplete septa.[2] A dense midline septum extending from the fetal neck across the full width of the hygroma is found. The septum represents the nuchal ligament.[5]

Fetal cystic hygroma colli requires prenatal diagnosis for proper management. The objective of this report is to present the characteristics of the ultrasound findings and neonates.

Materials and Methods

Ultrasonographic examinations were performed by the authors from June 1989 to July 1995, using convex transducers (Aloka Model 650 or 680). Indications for ultrasonographic examinations included abnormal growth, threatened abortion, suspicion of fetal death, etc. The most important sonographic findings were cystic masses at the posterolateral aspects of the neck and the absence of spinal dysraphism or calvarial defect. Other associated anomalies were also carefully identified and documented.

When cystic hygroma was diagnosed, previous obstetric history was carefully reviewed and the counseling was given. The patient was followed until discharged from clinic.

Table 1. Baseline characteristics of the patients

No.	Age (years)	Parity	Gestation at diagnosis (weeks)	Indications for ultrasound examinations
1	30	0-0-0-0	15	Threatened abortion
2	32	0-0-0-0	16	Threatened abortion
3	24	2-0-0-2	19	Suspected fetal death
4	19	1-1-0-1	11	Threatened abortion
5	25	1-0-2-1	18	Small-for-date
6	24	0-0-0-0	12	Threatened abortion
7	36	0-1-1-1	16	Amniocentesis for genetic study
8	35	3-0-1-3	15	Large-for-date
9	20	0-0-0-0	20	Uncertain date
10	23	0-0-0-0	18	Threatened abortion
11	24	0-0-0-0	19	Large-for-date
12	38	1-0-0-1	16	Small-for-date

Table 2. Ultrasound findings and chromosome studies

No.	Ultrasound findings Cystic Hygroma	Associated Findings	Chromosome Studies
1	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck extending to axilla and mediastinum	-oligohydramnios -hydrops fetalis	Not done
2	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck	-	46, XY
3	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck extending to axilla and mediastinum	-oligohydramnios	45, XO
4	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck	-hydrops fetalis	46, XX
5	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck extending to axilla and mediastinum	-oligohydramnios -hydrops fetalis	Not done
6	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck	-	Not done
7	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck extending to axilla and mediastinum	-oligohydramnios -hydrops fetalis	47, XX ; + 21
8	-two locculations thick-walled -midline septation at posterior, posterolateral aspects of neck	-polyhydramnios -hydrops fetalis	46, XY

No.	Ultrasound findings Cystic Hygroma	Findings Associated	Chromosome Studies
9	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck extending to axilla and mediastinum	-oligohydramnios -hydronephrosis	Not done
10	-asymmetric multiseptate thin-walled -midline septation at posterior, -posterolateral aspects of neck	-	46, XX
11	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck	-oligohydramnios -marked hydrops fetalis -ventriculomegaly	45, XO
12	-asymmetric multiseptate thin-walled -midline septation at posterior, posterolateral aspects of neck	-marked hydrops fetalis	Not done

Results

Twelve cases of cystic hygroma colli were diagnosed and followed by the authors. The diagnosis of all cases was postnatally confirmed by autopsy. The demographic informations and detailed ultrasound findings are presented in Table 1 and 2 respectively. The majority of cases were complicated with threatened abortion (5 cases). Two patients, however, presented as large for date (one had polyhydramnios and the other with severe hydrops).

The maternal age ranged from 19 to 38 years, the mean age was 27.50 ± 6.45 years, 50% of the patients were primigravida.

All of them manifests as unexpected findings on the antenatal ultrasound with various indications during first half of pregnancies. The mean gestational age at time of diagnosis is 16.25 ± 2.77 weeks, range 11-20 weeks.

All of twelve cases were sonographically diagnosed in the first half of their pregnancies with no false positive. A dense midline septum could be identified in all cases. The majority of cases showed asymmetric, thin-walled, multiseptate cystic masses at posterolateral aspects of the neck. In 5 cases, cystic hygroma extended into the axilla or mediastinum. Of 12 cases, 7 had already developed some degree of hydrops fetalis (2 cases were severe). Fifty-eight percent (7 in 12 cases) had oligohydramnios and only one case had polyhydramnios. All of them had undergone elective termination of pregnancy and autopsies were performed. Three of 7 fetuses which chromosome studies were done had chromosomal abnormalities, 45 XO in 2 cases and trisomy 21 in 1 case. Excluding hydrops fetalis, other

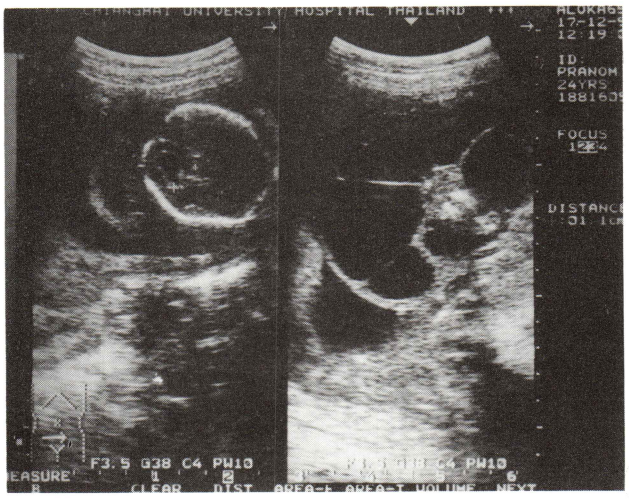

Fig. 1. Left : Cross section of the skull at the level of lateral ventricles shows ventriculomegaly and marked scalp edema.

Right : Cross section of the neck shows multisaptate thin-walled cystic hygroma.

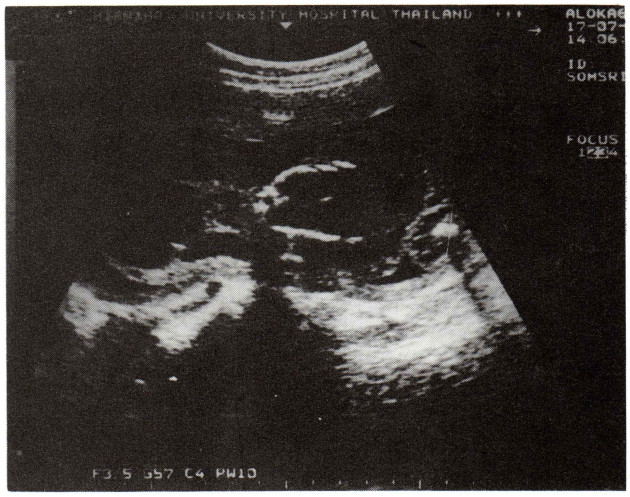

Fig. 2. Multiseptate thin-walled cystic hygroma at the posterior aspects of the neck (case 2).

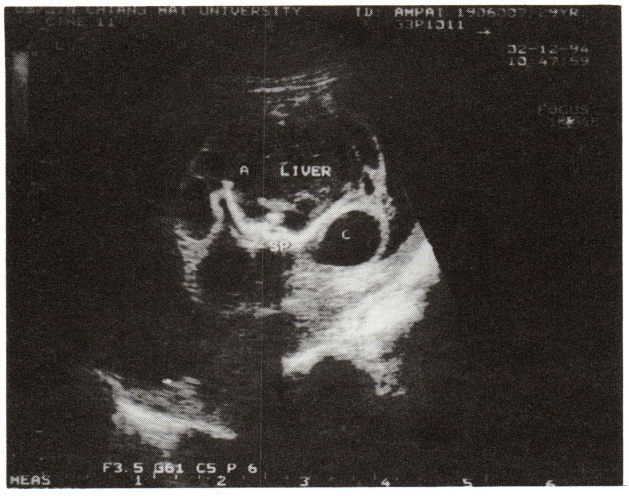

Fig. 3. Cross section of fetal abdomen shows multiseptate cystic hygroma extends to the level of abdomen (A = ascites, sp = spine, c = cystic hygroma).

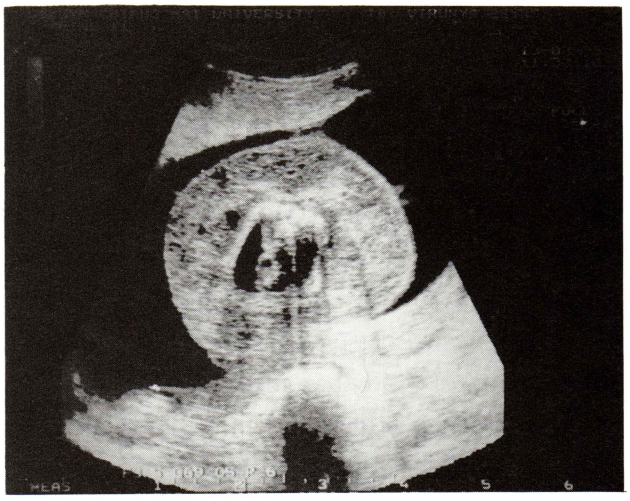

Fig. 4. Cross section of fetal chest shows marked subcutaneous edema of the fetus (hydrops fetalis).

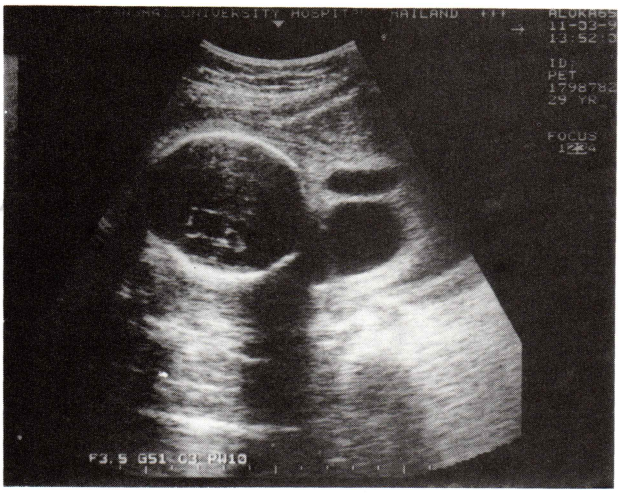

Fig. 5. Cross section of the skull shows cystic mass with thick wall at the posterior aspect of the skull (case 8).

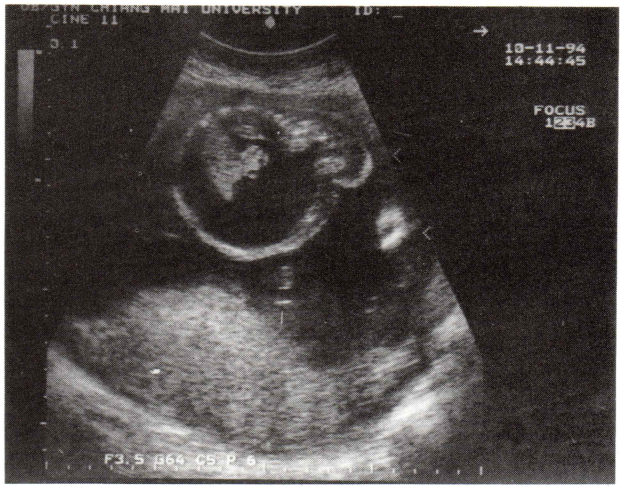

Fig. 6. Cross section of fetal abdomen shows marked ascites (case 10).

detectable anomalies included hydronephrosis (1 case) and ventriculomegaly (1 case).

Discussion

The sonographic findings of 12 cases of cystic hygroma in this study include asymmetric, thin-walled multiseptate, cystic masses without solid components at the posterolateral aspects of the neck. Interestingly, thick-walled cyst was found in one case (case no. 8 ; figure 5) and the cyst in this case had only two loculations and similar to cervical or low occipital meningocele. However, identification of the characteristic nuchal ligament and the absence of dysraphism or calvarial defect discriminates cystic hygroma from meningocele.

Similar to other reports,[3-6] oligohydramnios complicates 60% of cases whereas polyhydramnios was found in only one case. Although polyhydramnios may occur in cystic hygroma, the sonologist must not misdiagnose large fluid-filled loculations of cystic hygroma for polyhydramnios. The aetiology for oligohydramnios remains conjectural but may possibly result from fetal hypoperfusion leading to decreased renal output ; the polyhydramnios probably represents a manifestation of hydrops rather than esophageal compression. Given the common association between cystic hygroma and hydrops, the discordantly low incidence of polyhydramnios and high incidence of oligohydramnios suggests that factors leading to oligohydramnios predominate in the vast majority of cases.

Excluding hydrops fetalis and aberrations of amniotic fluid volume, other detectable anomalies associated with cystic hygroma were uncommon. We found only one case of hydronephrosis and and another case of mild ventriculomegaly.

Similar to other report,[6] midline septation extending from the posterior neck representing the nuchal ligament outlined by bilateral cysts were identified in all cases and may be the most specific sign of cystic hygroma.

Unfortunately, chromosomal study was carried out in 7 cases in this series, and only 3 cases or 43% were found to be abnormal. This is somewhat lower than in other reported series

(47-65%).[7-9] However, our series is too small to make a definite conclusion. Monosomy X was the most frequently reported karyotypic abnormality associated with fetal cystic hygroma, but trisomy also often encountered with cystic hygroma.

Because the majority of reported cases had undergone elective termination of pregnancy, the mortality rate of cystic hygroma diagnosed prenatally can not be stated with certainty. However, a compilation of small series and case reports of fetuses not electively terminated suggests a spontaneous mortalitiy rate of approximately 80-90%. The presence of hydrops fetalis or lymphangiectasia in cystic hygroma portends a grave prognosis, death usually occurring at several weeks from the time of diagnosis.[5]

Once a cystic hygroma is detected, a careful search is made for associated skin edema, ascites, and pleural or pericardial effusions. The outcome of fetuses with cystic hygromas varies but can result in intrauterine demise or partial regression, leaving a webbed neck. Rarely, they may be localized with fairly normal outcome.

In conclusion, this small series indicates that ultrasound examination of cystic hygroma typically shows asymmetric, thin-walled, multisepate, cystic masses at the posterolateral aspect of the neck but may extend into the axilla or mediastinum. Ultrasound had high predictive value in diagnosis of cystic hygroma. The common associated abnormalities were hydrops fetalis and oligohydramnios.

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