

Fetal Cystic Hygroma

FETAL KİSTİK HİGROMA*

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SUMMARY

A case ofprenatally diagnosedfetal cystic hygroma detected during ultrasound examination is reported. Fetal nuchal cystic hygroma is a congenital malformation of the lymphatic system appearing as a single or multinucleatedfluid-filled cavities which usually effect the postnuchal region. It arises as a consequence of a lag in the normalformation of a communication between the developing jugular lymph sacs and the internal jugular vein. An ultrasound diagnosis of cystic hygroma should be followed by a careful search for other anomalies and by fetal karyotyping. In this case, the cause and the natural history of this malformation has been discussed additionally.

KeyWords: Felal cystic hygroma

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Fetal cystic hygroma is a malformation consisting of one or more fluid filled lymphatic spaces in the posterior neck. Synonyms include; posterior nuchal cystic hygroma,fetal cystic lymphangioma, posterior cervical cystic hygroma, cervical lymphocele, hygroma cervicis, jugular lymphatic obstruction sequence and nuchal bleb. It is usually an isolated

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ÖZET

Ultrasonografik muayene sırasında prenatal olarak tesbit edilmiş bir fetal kistik higroma olgusu sunulmuştur. Fetal kistik higroma, genellikle boynun arka bölgesini etkileyen, tek veya birçok sıvı-dolu kavite şeklinde gözlenen lenfatik sistemin konjenital bir malformasyonudur. Bu oluşum gelişmekte olan jugular lenf torbalarıyla internal jugular ven arasındaki bağlantının normal formasyonundaki bir gecikme sonucunda meydana gelir. Ultrasonografik olarak konulan kistik higroma tanısını, diğer anomalilerin dikkatli bir şekilde taranması ve fetal karyotip araştırması izlenmelidir. Bu vakada, bu malformasyonun sebebi ve doğal seyride ilave olarak tartışılmıştır.

Anahtar Kelimeler: Fetal kistik higroma

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lesion without syndromic associations. Several syndromes such as Turner, Noonan, Achondroplasia, Lethal multipl pterygium, Roberts, Gumming, Cowchock syndromes and teratogenic exposures such as alcohol, aminopterin and trimethadione has been reported in association with cystic hygroma (1-8).

CASE REPORT

The mother is 30 year old, gravida 2, para 1, healthy woman whose first child was delivered near term vaginally. The child is healthy. During her follow up, she underwent an ultrasound examination at about 30 th week of her gestation. Using a commercial linear scanning system (Hitachi EUB-200) a single active fetus with regular heart rate (142/min.) was found. The fetus was in cephalic presentation.

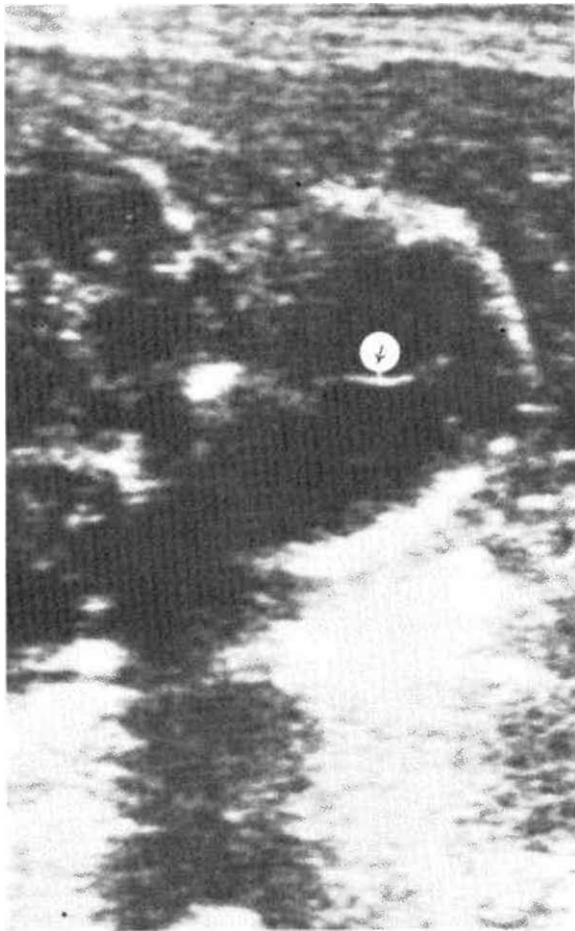


Figure 1. Ultrasound picture of the neck region of fetus showing fluid-filled area with a midline septum (arrow).

The placenta was implanted in fundal region and amniotic fluid volume was normal (amniotic fluid index = 12).

Further inspection revealed a bilobed, smooth-walled cystic mass projecting posteriorly from the region of the occiput (Fig. 1). The cranium appeared intact, the lateral ventricles were normal in size and location. Horizontal cranial section along the occiputofrontal axis showed a delicate septum (arrow) extending dorsally from the soft tissues of the neck defines a bilobed cystic mass. The cervical, thoracic and lumbar spine and long bones of the limbs appeared normal. The 80 mm. biparietal and 55 mm. femoral diameters corresponded to the 30-weeks gestational age.

After being discussed appropriately, this case was supposed to be an isolated cystic hygroma. After the natural course of the pregnancy a cesarean sec-

tion had to be performed because of the rupture of the membranes at 36 weeks gestation. The gross anatomic findings of the neonate weighed 3300 grams were as predicted by ultrasound examination with a bilocular, cystic structure projecting from the dorsal aspect of the neck (Fig. 2). Postnatal chromosome analysis revealed a normal male.

COMMENT

Fetal cystic hygromas have been explained pathophysiologically as a manifestation of the jugular lymphatic obstruction sequence (1-4). The fetal lymphatic vessels drain into two large sacs lateral to the jugular veins. These jugular lymph sacs eventually form communications with the venous system and become the terminal portions of the right lymphatic duct and thoracic duct. If lymphatic and venous structures fail to connect, the jugular lymph sacs



Figure 2. Newborn with a nuchal cystic hygroma

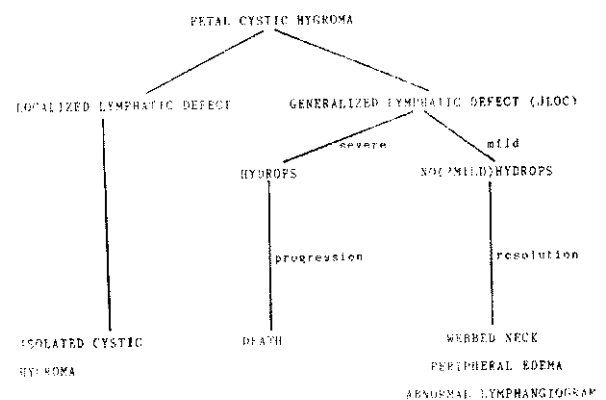


Figure 3. Natural History of Fetal Nuchal Cystic Hygroma A generalized lymphatic defect results from the jugular lymphatic-obstruction sequence (JLOS). (From Chervenak FA, Isaacson G, Blakemore KJ, et al: Fetal Cystic hygroma: Cause and natural history. N Engl Med 309: 322, 1983)

enlarge and lymph accumulates in tissues (2-4). The natural history of fetal cystic hygroma can be seen in (Fig. 3) (3).

In our case the cystic hygroma was discovered unexpectedly. It is evident that consistent diagnosis of this lesion requires routine sonographic examination of the fetal neck. An ultrasound diagnosis of cystic hygroma should be followed by a careful search for other anomalies and by fetal karyotyping (2,4).

The differential diagnosis of cystic of hygroma should include meningocele, benign cystic teratomas, nuchal edema, encephalocele and subchorial plasental cyst. The diagnosis should therefore be based upon at least two bilaterally echo-free symmetrical areas in the postnuchal region usually divided by septa and a completely formed cranical vault and a constant location of the masses with respect to the occiput even during fetal motion (1,3,4,5).

If hygroma is associated with hydrops, the chance of survival is small. There is scant information, however, about prognosis in the absence of hydrops but the ultimate prognosis of cystic hygroma is poor and pregnancy termination should be seriously considered in all prenatally diagnosed cases (4).

It has been thought that cystic hygroma colli was diagnostic of Turner's syndrome in utero but collected data demonstrate that Turner's syndrome is not the only cause of cystic hygromas (7). Already, several single-gene disorders and teratologic exposures have been associated with these malfor-

mations. In the cases of chromosomal abnormalities such as monosomy and trisomy, the recurrence risk will be very low, when but normal karyotypes are found one must consider non-chromosomal syndromes which have a recurrence risk up to 25% (2,3,4,7). Therefore monitoring of future pregnancies with ultrasound and possibly with fetal karyotyping should be offered to the family.

REFERENCES

1. Rottem S, Chervenak FA. Ultrasound diagnosis of fetal anomalies. Antepartal fetal surveillance. *Obstetrics and Gynecology Clinics of North America* 1990; 17: 17-40.
2. Elejalde BR, Elejakle MM, Leño J. Nuchal Cysts Syndromes. Etiology, Pathogenesis, and prenatal diagnosis. *AmJMedGen* 1985;21:417-32.
3. Chervenak FA, Isaacson G, Blakemore KJ, et al. Fetal cystic hygroma-Cause and natural history. *N Engl J Med* 1983; 309: 822-5.
4. Edwards MJ, Graham JM. Posterior nuchal cystic hygroma: Fetal Dysmorphology. *Clinics in Perinatology* 1990; 17: 611-40.
5. Garden AS, Benzie RJ, Miskin M, et al. Fetal cystic hygroma colli: Antenatal diagnosis significance and management. *Am J Obstet Gynecol* 1986; 154: 221-5.
6. Pijpers L, Reuss A, Stewart PA, et al. Fetal cystic hygroma: Prenatal diagnosis and management. *Obstet Gynecol* 1988; 72:223-4.
7. Carr RF, Oclis RH, Ritter DA, et al. Fetal cystic hygroma and Turner's syndrome. *AJDC* 1986; 140: 580-3.
8. Frigoletto FD, Binrholz JC, Driscoll SG, et al. Ultrasound diagnosis of cystic hygroma. *Am J Obstet Gynecol* 1980; 7: 962-4.