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## Abstract:

Cystic hygroma or cystic lymphangioma is a rare developmental congenital lymphatic malformation. It is most commonly related to thorax and neck region but it can also involve axilla, mediastinum, limbs and face. It is characterized by multiloculated cystic lesion of variable sizes. Ultrasound is very important in the diagnosis of prenatal cystic hygroma but it is also important in the characterization and differential diagnosis of cystic hygroma postnatally. In radiology department CT and MRI is done for the differentiation of cystic hygroma from other cystic lesions but due to the hazardous effects of these modalities it is important to give preference to ultrasound in the diagnosis of cystic lymphangioma. Some international studies are available on the cystic hygroma of the axillary region in infants but no local studies are published on the role of ultrasound in the diagnosis of cystic hygroma in infants.

## **Objective:**

To Determine Infantile Axillary Cystic Hygroma through ultrasonography.

## **Methods:**

We present a case of axillary cystic hygroma in infant of 3 months, diagnosed with ultrasound. An infant was brought to Gilani Ultrasound Center by his grandmother with a mass in the right axillary region, to search for its possible cause and extension to the adjacent organs. We diagnosed it as cystic hygroma with certainty by Ultrasound. The result was later on confirmed by surgery in children Hospital Lahore.

#### **Conclusions:**

Ultrasound is more important in the

characterization and diagnosis of cystic hygroma in infants.

## **Keywords:**

Cystic hygroma, Cystic lymphangioma, Axillary CH, Infantile cystic hygroma, Macrocystic lesion, septated non-septated cystic hygroma.

### Introduction:

Cystic hygroma of the infants is a rare congenital malformation of the lymphatic system. This macrocystic developmental lymphatic malformation, is characterized by fluid-filled sac that results from a blockage in the lymphatic system. A cystic hygroma may be fetal cystic hygroma; and discover during pregnancy with ultrasound, congenital; occurs as a birth defect in a newborn as a large, soft mass covered by normal skin, or it may not become evident until later in life <sup>1</sup>. It is highly associated with chromosomal abnormalities such as turner syndrome trisomy 13, trisomy 18 and down syndrome. It could be inherited in the form of syndrome (having multiple characteristic) or isolated. Isolated cystic hygroma could be inherited as autosomal recessive characteristic. Some researches while correlating the ultrasound results with amniocentesis estimated that cystic hygroma is more related to chromosomal abnormalities as compared to nuchal thickening<sup>2</sup>. Cystic hygroma with chromosomal association have poor outcomes. A study describe the association of cystic hygroma chromosomal abnormalities estimated 9% of cases occur in healthy infants, while 2% occur in liveborn

but with chromosome abnormalities or various malformations<sup>3</sup>.

It is believed that it is the abnormal junction of the lymphatic channels with the jugular vein. As evident from various sonographic appearances that abnormal transformation of lymphatic fluid in the jugular lymph sacs occurs. It is also believed that malorientation and blockage in the lymphatic vessels results in the cystic lymphangioma<sup>4</sup>. If the junction between the venous system and the jugular lymphatic sacs is not established, the lymphatic fluid will continuously grove and ultimately will result in subcutaneous edema, pleural or pericardial effusion and fluid will also be accumulated in the abdominal potential spaces in the form of ascites. If the connection is reestablished surgically or by any other invasive procedure the distended lymphatic sacs will gradually subside<sup>5</sup>.

#### Case:

A male infant of 4 months is sent to the Gilani ultrasound Center for the diagnosis of a large soft lump in the axillary region. The mass was nonpulsatile, soft and was looking non-tender because the infant was not showing worsening response to compression. It was first baby; no history of Rh incompatibility was known. No history of maternal hypertension, diabetes or any other chronic disease.

#### **Presentation:**

The baby was examined with Toshiba Xario with linear transducer 7-14MHz and convex transducer 3-6 MHz. Color, power and spectral Doppler were applied to know about blood flow in the mass, feeding vessels or any communication with the blood vessels. The procedure was performed under the AIUM practice guidelines, which are routinely observed in this department. The procedures were explained to the baby's grandmother and informed consent was signed. Preapproval was obtained from the institutional review board (IRB) and ethical committee. A large multiloculated cystic lesion was observed with septation, it was large enough cystic lesion to be occupied in the field of view of the linear transducer (Figure 1). For the visualization of its global view convex array transducer with 6cm footprint transducer was selected and the mass is measured, dimensionally it was 51.1x43.5x48.4mm, which is equal to  $56.3cm^3$ (Figure 2). No blood flow was detected on color Doppler, although acoustic streaming was seen with compression (Figure 3 & 4). There was no communication of the cystic lesion with any vessel or lungs. It was outside the ribcage, just beneath the skin, in the subcutaneous region. All the sonographic appearances and Doppler criteria was suggestive of cystic hygroma (cystic lymphangioma).



Figure 1: Baby of 4months having large, Soft, non-pulsatile and non-tender mass at the right axillary region.



Figure 2: Cystic hygroma seen with linear transducer



Figure 3: Cystic Hygroma 51.1×43.5×48.4mm



Figure 4: Color Doppler image with acoustic streaming vascular

The result was later on confirmed by surgery in children Hospital Lahore. The grandmother of the infant was put on the call to know about the case. She with thanks that the surgery have been conducted and confirmed that the ultrasound diagnose was accurate, the case was cystic hygroma.



Figure 5: Color Doppler applied through compression

#### **Discussion:**

Cystic hygroma of the axillary region is an uncommon, rare but possible condition especially in newborn and infants. We examined a baby with large palpable, soft, non-pulsatile and nonpalpable mass in the right axillary region extending from right nipple to occupy all of the armpit. Cystic hygroma is a lymphatic system malformation; it may either be complete blockage (non-communicated) or temporary block of the lymphatic system. If the jugular lymphatic sacs are totally non-communicated with the venous system then it will result in septated, multiloculated, mega-cystic hygroma, which is not evident to regress spontaneously. But unilocular cystic hygroma is believed to be due to partial obstruction of the lymphatic system and observed to resolve spontaneously. It is believed that increase in the hydrostatic pressure of cystic hygroma sometimes overcome the partial obstruction and thus spontaneous resolution of these lesions<sup>6</sup>.

Cystic lymphangioma is found to be associated sometimes with fetal hydrops, aneuploidy, intrauterine fetal death and structural malformations <sup>7</sup>. Infantile cystic hygroma is estimated from the known data of a demographic research from 1/ 6000 to 1/ 16,000 but the actual occurrence will be much more that due to unavailability of data on one hand but on the other hand this is an old information <sup>3</sup>.

The report of a study regarding cystic hygroma was published in 2007, they studied a population of 38167. They found 134 (1 in 285) as cystic lymphangioma. They found 51% cystic hygroma were associated with chromosomal abnormalities most commonly 18.65% trisomy-21, 14.17% Turner syndrome, 9.7% trisomy18. Cardiac and skeletal malformation were diagnosed in about 34%. Overall, 17% was estimated survival<sup>8</sup>. Another study was conducted on 72 cases of cystic hygroma and 4(52.7%) associated with chromosomal abnormalities. 18 infants born alive, they were followed for 2 to 8 years. 1 developed Noonan's

syndrome and 1 had problem in urinary system while 16 found normal. It is obvious from the data that prognosis of the cystic hygroma has very poor prognosis in the early days of development<sup>9</sup>.

Cystic hygroma carries risk for chromosomal abnormalities and fetal structural malformations. Fetuses diagnosed with cystic lymphangioma should be followed with ultrasound antenatally for other structural or chromosomal abnormalities. karyotyping should be advised to rule out possibilities of chromosomal abnormalities. Fetus should be monitored for fetal hydrops with series scheduled antenatal ultrasound. For the prognostic perspective, the lesion should be monitored for size, structure and septation. Spontaneous resolution, less or no septas and the axillary location of the lesion are regarded as good prognostic factors<sup>1014</sup>.

## **Conclusios:**

Ultrasound is the modality of choice for the diagnosis and characterization of any soft tissue cystic or solid lesion and especially cystic hygroma, pre-or postnatally.

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