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**Radiological evaluation of a large fetal cystic hygroma**

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**ABSTRACT**


A routine antenatal ultrasonography at 20 weeks of gestation revealed a large multicystic mass lesion arising from the neck region. The lower half of the face, mandible and lips could not be differentiated from the lesion. An MRI was performed to see the extent of the lesion. It revealed a large well defined fluid intensity lesion arising from the oral cavity and anterior aspect of neck, extending upto the xiphisternum externally. Internally it was seen compressing the oesophagus and larynx with intrathoracic extension into the mediastinum and abutting the heart. The patient was followed postnatally when a full term live baby was delivered by lower segment caesarean section which succumbed few hours later. Our findings were confirmed and clinically relevant photographs were obtained. Cystic hygroma or nuchal lymphangioma arises from delayed development or maldevelopment of the lymphatic system to communicate with the venous system of the neck.

**Keywords:** Nuchal translucency, Lymphangioma, Cystic hygroma, Congenital anomaly, Fetal/Antenatal ultrasound.

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## INTRODUCTION

Cystic hygroma or nuchal lymphangioma are rare, benign cystic lesions of vascular origin. They show lymphatic differentiation and are considered the lymphatic equivalents of haemangiomas of blood vessels [1] [2]. They arise from delayed development/maldevelopment/failure of the lymphatic system to communicate with the venous system of the neck. They can occur at almost any location and have a marked predilection in the head and neck i.e. 95% in the neck and axillary regions, where they are also called nuchal cystic hygroma. Other locations include the mesentery, retroperitoneum, abdominal viscera, lung, and mediastinum. Few other rare ones include hepatic, splenic, renal and pancreatic lymphangiomas [3].

Only 10% of them are known to extend into the mediastinum. Only 0.2-0.3% of them is seen in the fetal population, out of which only 1 in 8000 pregnancies results in live births [4], thus making this case a rare one. We present to you this case of a large cystic hygroma with widespread extensions, diagnosed using ultrasound and MRI with the surgical pictures. It needs to be stressed further that in large lesions like these, ultrasound is insufficient to describe its extent and fetal MRI is advisable.

## MATERIALS AND METHODS

Ultrasound was performed using a Mindray DC-7, MX-26003711 machine, curvilinear probe and the required images stored. The Magnetic resonance imaging was performed using a 1.5 Tesla GE machine. The delivery took place at our tertiary care institute using routine lower segment caesarean section delivery protocol.

## RESULTS

A 31 year old female came for routine antenatal ultrasonography at 20 weeks of gestation (19 weeks 6 days by last menstrual period and 20 weeks by first scan). Due to economic constraints and lack of availability of radiologists in her village, this was unfortunately her first scan.

Ultrasonography revealed a large well defined thin walled anechoic multicystic mass lesion arising from the neck region. The lower half of the face, mandible and lips could not be differentiated from the lesion (figures i to v). The brain, heart, abdominal organs and limbs, however appeared unremarkable. We followed up with the patient ultrasonographically every month. No evidence of any cardiac or skeletal abnormality was noted. The amniotic fluid index was within normal limits and no signs of hydrops fetalis development were seen.

As the patient had conceived after 4 years, she was anxious for a baby. However she could not afford any chromosomal tests. An MRI was performed to see the extent of the lesion. It revealed a large well defined altered signal intensity multi-septate cystic lesion involving the anterior aspect of the neck and measuring approximately 10.5 x 7.5 x 12.1 cm (Transverse x Anteroposterior x Craniocaudal). It appeared hypointense on T1W images and hyperintense on T2W and STIR sequences (figures vi to x).

The lesion was seen involving the oral cavity and extending into the anterior aspect of the neck. Inferiorly, there was a large exophytic component seen reaching upto the xiphisternum externally. The lower lip and mandible were not seen separately from the lesion. In the neck, it was seen compressing the proximal esophagus and larynx with resultant dilatation of the hypopharynx. It also showed focal intrathoracic extension into the antero-superior mediastinum, abutting the heart.

The patient was followed postnatally when a full term live baby was delivered by lower segment caesarean section which unfortunately succumbed after few hours. Our findings were confirmed and clinically relevant photographs obtained, following prior informed consent (figures xi, xii and xiii).

## DISCUSSION

Cystic Hygroma was first described in the European Literature and is also known as cystic lymphangioma and hygromacolliscysticum. The term hygroma means moist tumor. Cystic hygromas are congenital vasculo-lymphatic malformations that have no predilection for sex or race and no malignant potential. Typical cystic hygromas, when seen in paediatric or adult population, cause no symptoms unless they are large in size or surround and invade adjacent normal anatomic structures. In this situation, cystic hygromas may cause symptoms such as feeding problems or breathing difficulties [9].

They can present at any age but most often occur in the paediatric population (~90% in those less than 2 years old) [10]. They occur most commonly in the neck, which is also termed nuchal cystic hygroma (occurs in ~80% of cases) and axilla, with only 10% of cases extending to the mediastinum and only 1% confined to the chest [8].

**Subtypes:** There are classified according to the size of the lymphatic cavities. In order of increasing size, they are:

- simple microcystic or capillary lymphangioma
- macrocystic or cavernous lymphangioma
- cystic hygroma: cystic lymphangioma (seen in this case)
- other: lymphovascular malformation (e.g. venolymphatic malformation).

#### **Radiographic features:**

On prenatal ultrasound, they may present as a well circumscribed nuchal cyst with or without evidence of fetal anasarca/hydrops fetalis. Greater volumes are thought to correlate with increased karyotypic abnormality and poor fetal outcome. Features seen on antenatal ultrasound are:

- multilocular cystic mass
- internal septa of varying thickness -the presence of septations may indicate a poorer outcome.
- cystic contents: usually anechoic, heterogeneously hyperechoic if debris, lipid content, infection or haemorrhage
- Rarely, solid areas or mostly solid with cystic foci
- On colour Doppler: +/- arterial or venous flow in the septa

In the head and neck region, larger lesions tend to occupy more than one deep space, sandwiching between normal structures.

On CT, most lymphangiomas appear homogeneous and cystic on CT, but some appear inhomogeneous because of the presence of proteinaceous, fluid, blood, or fat components within the lesion. It is rare for CT to demonstrate intrinsic septations.

On MRI, signal characteristics include:

- T1: predominantly hypointense unless there are haemorrhagic components
- T2: predominantly hyperintense
- T1C+ Gd (in adult or pediatric populations, where contrast study is performed if required. Not done in fetal MRI): no enhancement except occasional faint enhancement of rim.

#### **Differential diagnosis**

Differential considerations on antenatal ultrasound include:

- cervical teratoma
- occipital encephalocele
- cervical meningocele

#### **Complications**

- Development of non-immune hydrops fetalis: which often indicates a poorer prognosis.

- respiratory obstruction from pharyngeal oedema in pediatric/ adult cystic hygroma

#### **Follow-up:**

- Follow-up scans every 4 weeks to assess the evolution of the hygroma and development of hydrops, if present.

#### **Delivery:**

- Place: hospital with neonatal intensive care and pediatric surgery.
- Time: 38 weeks, earlier if hydrops develops.
- Method: cesarean section if there is hydrops or large cystic hygromas preventing flexion of the head.

#### **Prognosis:**

- Fetal death: 90%.
- In 10% of cases the fetal karyotype is normal, there are no other obvious defects and the hygromas resolve during pregnancy. In these cases the prognosis is good.

#### **Recurrence:**

- Isolated cystic hygroma or as a part of Turner syndrome: no increased risk of recurrence.
- Part of autosomal recessive syndromes: 25% risk of recurrence.
- In this case, the same patient happened to come back to us 14 months later with a second pregnancy! We have, at the time of submitting the article, performed four antenatal ultrasound scans on her- at 8 weeks (dating scan), 11 weeks (NT scan), 19 weeks (anomaly scan) and 29 weeks gestation. The fetus appears normal with no detectable malformations detected.

## **CONCLUSION**

A detailed antenatal scan is extremely essential and in large lesions where ultrasound is insufficient to describe the extent, fetal MRI is always advisable. Antenatal cystic hygroma is a benign but invasive appearing lesion which usually has a poor prognosis. It is the cause of a lot of emotional trauma to the parents and the family, hence it's early detection and thorough counseling is a must. Congenital anomalies are a significant cause of disability, chronic illness, and childhood death in many countries affecting approximately 1 in 33 infants. They result in an estimated 3.2 million birth defect-related disabilities every year. 2-3% of all births are complicated by congenital anomalies and therefore, they are an important cause of perinatal morbidity and mortality accounting for

20–30% of perinatal deaths [Quoted from Congenital anomalies fact sheet: WHO, 2015]. The early detection of foetal anomalies is beneficial for both high and low risk groups. A negative sonogram may be reassuring, especially for couples at increased risk. On the other hand, positive

findings enable decisions in early gestation when termination of pregnancy might be preferable. The reported case has images of ultrasound, MRI as well as clinical images post-delivery, thus serving as a comprehensive pictorial essay in understanding the condition.

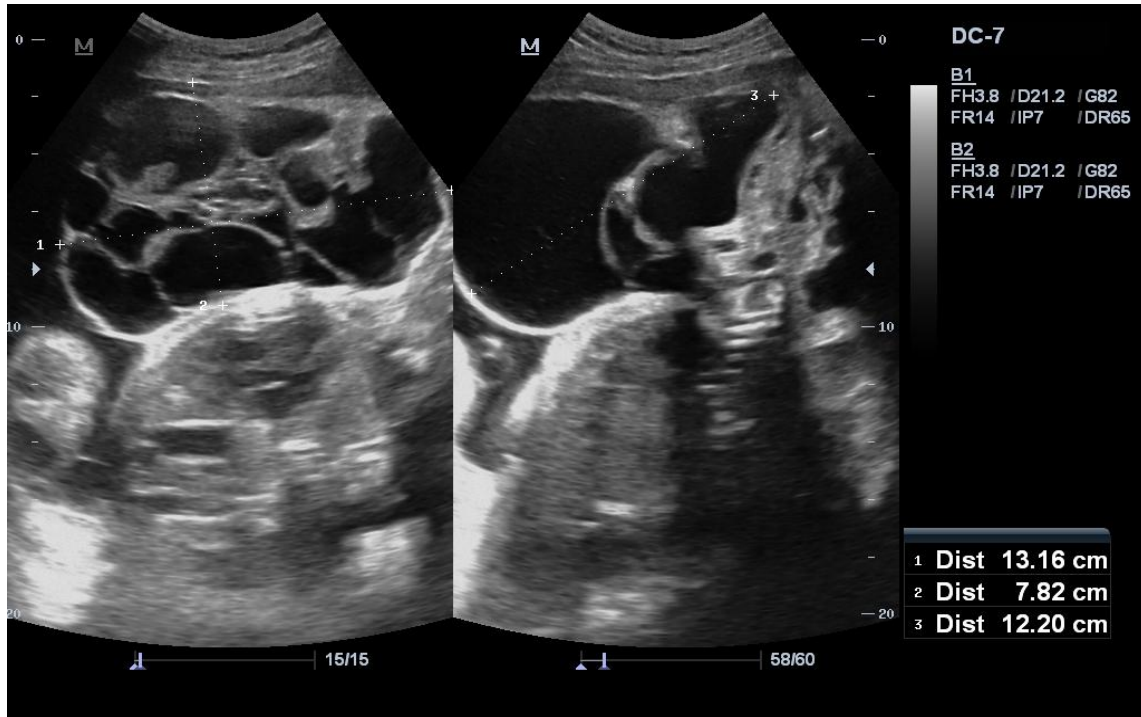


Figure i: ultrasound image showing large well defined thin walled multicystic lesion.

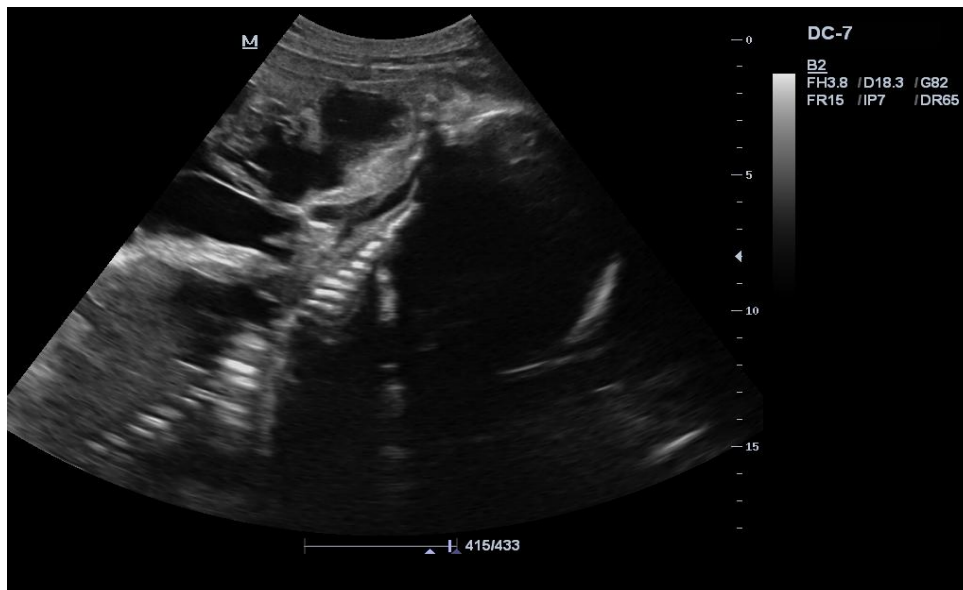
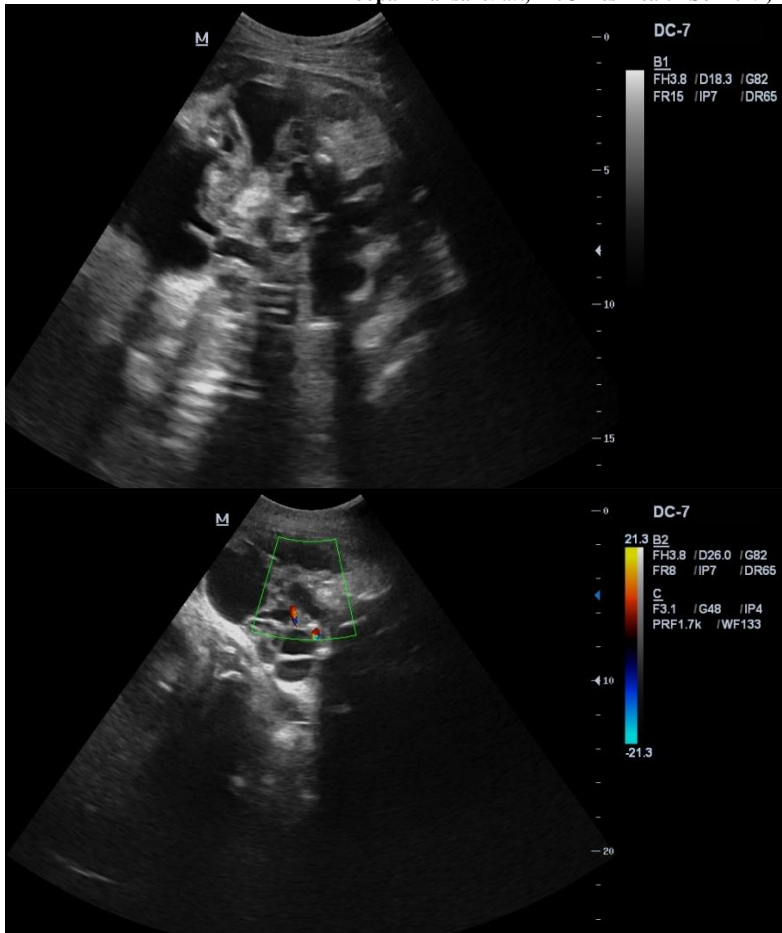


Figure ii: ultrasound image showing location of the lesion, adjacent to the vertebral column, from the neck region.



Figures iii and iv: ultrasound images showing the lesion arising from the neck region and showing no vascularity within.

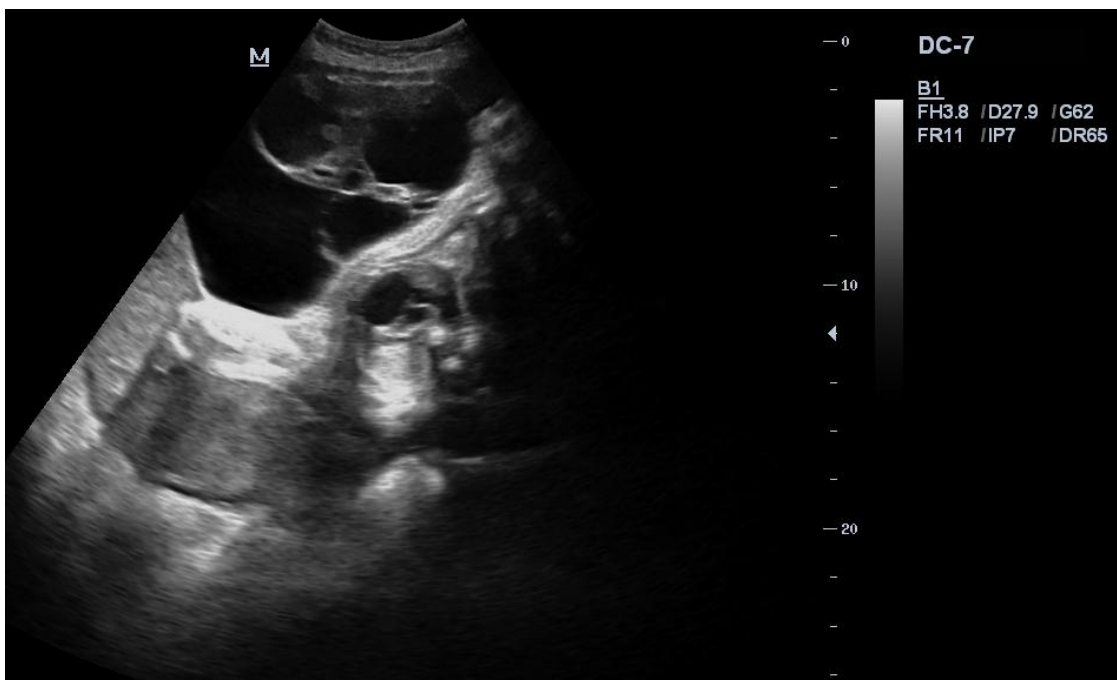
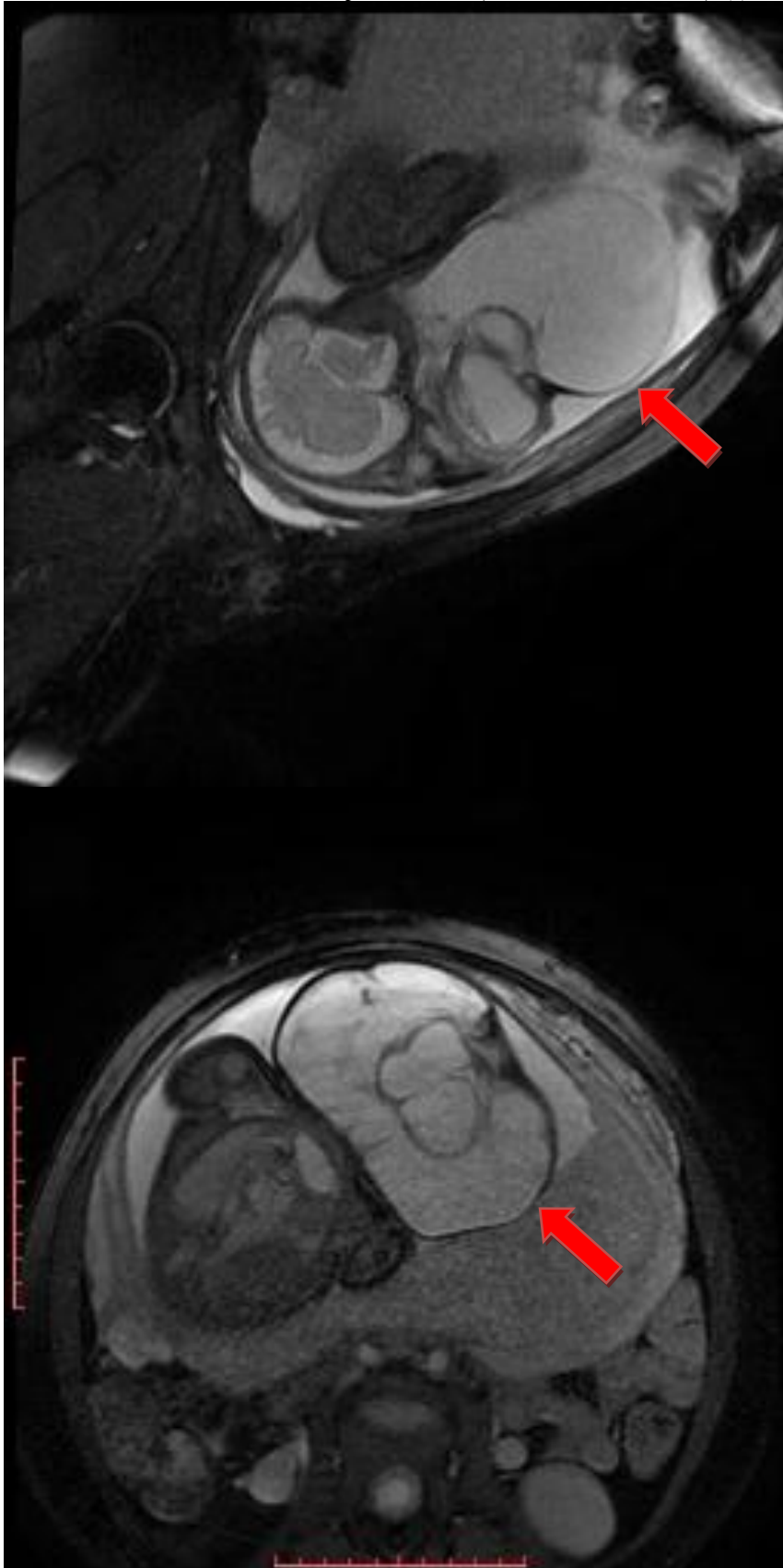
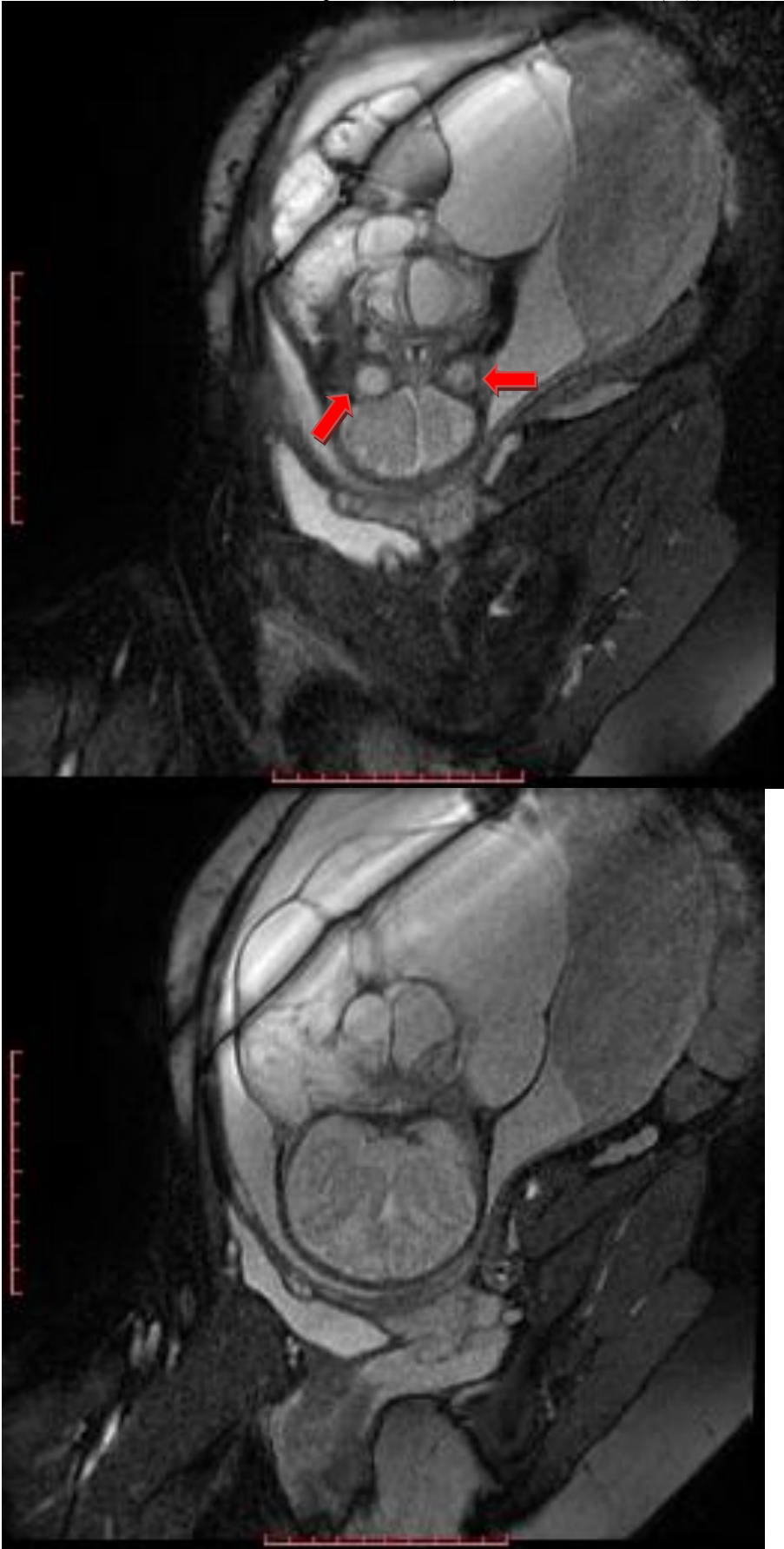


Figure v: ultrasound image showing lesion separate from the uterine wall and the placenta.



Figures vi and vii: MRI sagittal and axial images showing the large fluid intensity multicystic lesion (arrows).





Figures viii, ix and x: MRI coronal and sagittal images showing the lesion adjacent to, but separate from the brain and upper face (eyes seen separately- arrows).







**Figures xi, xii and xiii: post LSCS images showing live born full term baby with large lymphangioma arising from the neck region.**

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