

The Value of Fetal Magnetic Resonance Imaging in Giant Lymphangioma: A Case Report

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ABSTRACT

A giant lymphangioma at the neck, axillary, chest wall up to abdominal wall, and extension into upper thoracic cavity and the right upper extremity is reported. Ultrasound examination during antenatal care revealed huge cyst lesion with septation. Magnetic resonance imaging was performed at 36 weeks of gestation to determine tissue characteristic, evaluate tumor extension especially into thoracic chest cavity for looking any compression vital structure such as airways, vascular, therefore can prepare of technique delivery of the baby and surgical planning after birth as well. Tracheoesophageal displacement index (TEDI) was measured more than 12. Fetal karyotype shows male karyotype with double deletion on chromosome 17. The baby has been operated 3 times. Histopathological result was lymphangioma. Intralesional bleomycin of 0.5 mg/kg is given post operation.

Key words: Abdominal wall, axillary, chest wall, giant lymphangioma, magnetic resonance imaging, neck, tracheoesophageal displacement index, thoracic cavity, ultrasonography, upper extremity

INTRODUCTION

ymphangiomas are congenital lymphatic malformations that are usually discovered at early childhood. Since the increase in routine prenatal ultrasonography (USG), the majority of cases can be diagnosed earlier. The incidence of lymphangioma was reported as one in every 6000 live birth. Lymphangiomas can manifest in any location of the body in the developing lymphatic systems but occurred most commonly in the neck, head, and axilla.^[1-3] Although having the characteristics of benign mass and poses no serious threat of morbidity or mortality, massive lesion can sometimes cause life threat due to compression or infiltration of adjacent structure such as the aorta and trachea. They have no malignant potential and no predilection of sex or any race, but in some authors, has described a male predominant.^[4,5] Surgical excision is the treatment of choice with recurrence rates as high as 15% has been reported.^[6-8] The success of surgical intervention and outcome depends on the size,

location, and type of tumor. Location, extension of the lesion, and other associated abnormalities determine the prognosis of lymphangiomas.^[6] USG is still the first method of fetal examination for diagnosis lymphangiomas.^[9] Magnetic resonance imaging (MRI) with larger field of view and good contrast resolution can display detailed anatomic for evaluating the extension and characteristics of lymphangiomas and can also measure tracheoesophageal displacement index (TEDI) which is important to predict airway complication.^[10,11] The huge lymphangioma encompasses the neck, axillary, thoracicabdominal wall, extremity, and infiltration into upper thoracic cavity is infrequent, hence why we reported this case as well as to determine the value of fetal MRI in giant lymphangioma case.

CASE REPORT

A 34-year-old second gravid, primipara was referred to the department at 34 weeks' gestation due to large cystic

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multiseptated mass seen in the neck, axillary, chest wall up to abdominal wall detected by routine US. The mass displayed no color flow on Doppler study, and hence, the most likely diagnosis is a lymphangioma. The patient's family history and previous medical history were unremarkable. The first child is in normal condition and no consanguinity of the parent. Amniotic fluid volume and other structures were within normal limits. MR was performed at 36 weeks of gestation with 1.5 T (Siemens Magnetom Avanto, Erlangen, Germany) with ultrafast sequence technique without contrast media intravenous injection. The images were obtained in multiplanar planes of fetal thorax and abdomen. MR reveals huge heterogeneous mass with cystic component and septate with multiple fluid level at the right side of the neck, right axilla, right thoracic-abdominal wall, and right upper extremity, the size approximately $13 \times 14 \times 19.8$ cm. Based on characteristic tissue and location, the diagnosis lymphangioma was established. The mass infiltration into upper chest cavity displaced subclavian artery and surrounding carotid artery [Figure 1a-c]. The trachea is displaced to the opposite side and ventral. Caliber of trachea and esophagus is still good [Figure 2a and b]. TEDI measurement is >12 [Figure 3], but there is no complication polyhydramnios. There is no intra-abdominal lymphangioma and no mass extension into the floor of the mouth. The multidisciplinary team approach was assembled involving obstetrics, pediatric surgeon, ENT



Figure 1: (a) Coronal T2HASTE: Cystic mass with septation and multiple fluid level. (b) Axial T2 HASTE at the level lower chest. The mass involved the arm (arrow) did not cross contra laterally. (c) The mass is displacing the subclavian artery (arrow)



Figure 2: (a) T2 HASTE coronal: Intrathoracic mass invasion (arrow). (b) T2HASTE sagittal: No esophagus compression (arrow)

surgeon, and anesthetist counseled the patient and also mode of delivery and further management. Team advised an *ex utero* intrapartum treatment (EXIT) procedure and explained about risk and benefit of this procedure. Patient decided to refuse of EXIT procedure during delivery baby. The baby



Figure 3: Tracheoesophageal displacement index measurement ±17.8 mm



Figure 4: After delivery, giant mass was seen at the right neck, axilla, thoracic-abdominal wall and arm, but spontaneous respiration occurred without any intubation



Figure 5: Chromosome analysis reveals double deletion in chromosome 17



Figure 6: Histopathological examination from the chest shows the presence of large, endothelial lined vascular space containing lymphocytic aggregates, and hemorrhagic fluid



Figure 7: Baby condition after 3 times operation

was born with elective section cesarean (SC) with Apgar score 2/8 and spontaneous respiration [Figure 4]. Karyotype from umbilical cord was done and the result revealed double deletion on chromosome 17 [Figure 5]. Histopathology confirmed lymphangioma [Figure 6]. Postnatally, the first operation was neck debulking. It was done at 2 months after birth followed by abdominal thoracic incision at 4 months and arm incision at 6 months of age [Figure 7]. Intralesional bleomycin was given post operatively with the dose of 0.5 mg/kg.

DISCUSSION

A giant fetal lymphangioma which is covered the right neck, axillary, chest wall, abdominal wall and arm, and invasion in right upper thoracic cage is reported. Lymphangiomas can be diagnosed mainly by USG as the first method of choice in fetal examination. Computerized tomography has to avoid for further diagnosis due to radiation exposure in fetal age.^[12] The advantages of excellent tissue contrast visualization have enable MRI to confirm diagnosis or to negate USG findings. Differential diagnosis of congenital cystic neck mass includes germ cell tumor, vascular malformation, and thymic cyst, while cervical myelomeningocele and brachial

cleft cyst are uncommon.[10,13,14] Signal characteristic of lymphangioma on MRI is usually hyperintense on T2W with variable intensity on T1W depending on protein content.^[15] Fluid-fluid level can be seen in cases complicated by hemorrhage. Germ cell tumors have mixt solid and cystic signal intensity with calcification component and septation.^[10,16]

There is no consensus regarding technique of the delivery. Whether the baby should undergo SC delivery, can be normal or EXIT procedure, the morbidity and mortality are increased when airway compression occurs. It is the important to identify the airway in case with fetal neck mass. The predictors for airway distress are polyhydramnios. teratoma, and TEDI >12 near delivery.^[10] USG has a limited role in assessing the extension of the mass due to difficulty in visualizing of larynx and trachea. The secondary sign such as polyhydramnios, small or undetectable stomach, decreased swallowing, or tongue protrusion could help USG to predict an airway and esophageal obstruction by Suzuki et al.[17] Fetal MRI can delineate esophageal pouch in case suspected of esophageal obstruction. MRI is excellent to visualize trachea position; therefore, the displacement can be measured precisely to estimate the severity of airway obstruction. In this case, all the predictors of difficulty of airway are not fulfill except one, TEDI of more than 12. During cesarean delivery, to overcome suspected airway obstruction, as the therapeutic option of operating on placental support, EXIT procedure was advised. The principle of procedure is the control of uterine hypotonic to preserve uteroplacental circulation with neonatal anesthesia and thereby obtain additional time to establish an airway. This procedure necessitates a team to be assembled. Maternal complications such as intrapartum hemorrhage from placental abruption or postpartum hemorrhage from uterine hypotonic should be considered. However, the patient refused EXIT procedure. Finally, the baby boy was delivered with SC without EXIT procedure with Apgar score of 2 at 1 min and 8 at 10 min; baby weighted 4628 g and required no intubation.

As reported by Steigman *et al.*, cervical teratoma rather than lymphovenous malformation would more likely require airway intervention at birth.^[18]

The frequency of the chromosomal abnormality associated with lymphangiomas is as high as 78%. Lymphangioma discovered in utero may have a poor prognosis when the karyotype is abnormal with fetal hydrops. This case revealed male karyotype with double deletion on chromosome 17. This chromosomal abnormality can be derived from the parents or de novo due to cleavage error in reproduction system. According to Carr *et al.*, Turner syndrome is being the most common,^[19] other reported trisomy 21 and trisomy 18.^[20] However, significant clinical diversity is observed within the same population of patients with the same

chromosomal anomaly, and frequently, particular pattern of congenital malformation does not specify a specific genetic disorder.^[21]

Management of giant lymphangiomas is always done by surgical excisions. Since the mass is huge and involvement many place of the body, it is impossible to completely excise the lesion in a single operation. Neck debulking was done at 2 months after birth followed by abdominal thoracic incision at 4 months and arm incision at 6 months of age. Post-operative intralesional bleomycin of 0.5 mg/kg is administered.

SUMMARY

Lymphangioma is benign lymphatic malformation that occurs more commonly in the head, neck, and axilla. Small lymphangiomas can regress spontaneously, but in large lymphangiomas tent to infiltrative into surrounding structure, need a team with multidisciplinary background for the successful delivery, and prevent respiratory obstruction. Fetal MRI is a valuable auxiliary modality to USG in confirmation of diagnosis, to exclude intra-abdominal lymphangiomas and to determine of the extent of the lesion, those permitting a planned delivery. In this case, the measurement value of TEDI as a reference to predict the difficulty of breathing at birth cannot be solely predictor used, rather also determined by the type of tumor and polyhydramnios.

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