Congenital Lymphangioma of the Fetal Lower Limb: A Rare Case

Diki Riandi,[1] Donel S[2]
[1]Resident of Obstetrics and Gynecology Faculty of Medicine, University of Riau/ Arifin Achmad General Hospital, Pekanbaru, Indonesia
[2]Department of Obstetrics and Gynecology Faculty of Medicine, Maternal-Fetal Medicine Division, University of Riau/ Arifin Achmad General Hospital, Pekanbaru, Indonesia
Correpondence: Diki Riandi, Email: dikiriandi73@gmail.com

Abstract
Introduction: Lymphangioma is a rare congenital malformation caused by the failure of primitive lymphatic channels to connect with the venous system. Its incidence is approximately 1 in 6,000 pregnancies, with only 2% affecting the limbs. Prenatal diagnosis remains challenging, requiring advanced imaging techniques for accurate differentiation from other congenital masses.
Case Presentation: A 39-year-old woman, G6P4A1L4, was referred to Arifin Achmad Hospital at 25+6 weeks due to fetal lower limb enlargement and ascites. Ultrasound revealed a large multicystic mass on the fetal left thigh and buttock, measuring 10.2 × 7.8 × 6.4 cm. Initially suspected as sacrococcygeal teratoma, further evaluation suggested lymphangioma. The Doppler study indicated reduced vascularity. After thorough counseling and an ethics committee review involving obstetricians, neonatologists, and pediatric surgeons, the parents opted for termination. Labor was induced with misoprostol 50 mcg orally every 4 hours (two doses), leading to the delivery of a 1,400-gram male infant with an APGAR score of 3 and 0. The baby died 15 minutes post-birth. Maternal condition remained stable with normal vital signs and laboratory results post-delivery.
Discussion: Prenatal diagnosis of lymphangioma is difficult due to its resemblance to other congenital masses. Regular extremity screening is crucial for early detection. Postnatal histopathology confirmed lymphangioma, showing dilated lymphatic channels without solid components. This case highlights the importance of multidisciplinary management and serial imaging for prognosis refining prognosis.
Conclusion: Prenatal diagnosis is crucial for management planning. Serial ultrasound and MRI can refine prognosis. A multidisciplinary approach is essential for optimal care.

Key words: Lymphangioma, Lymphatic malformation, Prenatal diagnosis, Fetal limb mass, Perinatal management.

Limfangioma Kongenital pada Ekstremitas Bawah Janin: Kasus Langka

Abstrak
Pendahuluan: Limfangioma adalah malformasi kongenital langka yang disebabkan oleh kegagalan saluran limfatik primitif untuk terhubung dengan sistem vena. Insidensinya 1 dalam 6.000 kehamilan, dengan hanya 2% yang mengenai ekstremitas. Diagnosis prenatal tetap menjadi tantangan, memerlukan teknik pencitraan canggih untuk membedakannya secara akurat dari massa kongenital lainnya.
Presentasi Kasus: Seorang wanita berusia 39 tahun dengan G6P4A1H4 dirujuk ke Rumah Sakit Arifin Achmad pada usia kehamilan 25+6 minggu karena pembesaran ekstremitas bawah janin dan asites. Ultrasonografi menunjukkan massa multikistik besar pada paha kiri dan bokong janin, berukuran 10,2 × 7,8 × 6,4 cm. Awalnya dicurigai sebagai teratoma sakrokoksigeal, namun evaluasi lebih lanjut mengarah pada dugaan limfangioma. Studi Doppler menunjukkan vaskularisasi yang berkurang. Setelah konseling menyeluruh dan tinjauan oleh komite etik yang melibatkan dokter obstetri, neonatologi, dan bedah anak, orang tua memilih terminasi kehamilan. Persalinan diinduksi dengan misoprostol 50 mcg per oral setiap 4 jam (dua dosis). Induksi ini menghasilkan kelahiran bayi laki-laki seberat 1.400 gram dengan skor APGAR 3 dan 0. Bayi meninggal 15 menit setelah lahir. Kondisi ibu tetap stabil dengan tanda vital dan hasil laboratorium normal pascapersalinan.
Diskusi: Diagnosis prenatal limfangioma sulit karena kemiripannya dengan massa kongenital lainnya. Skrining ekstremitas secara rutin sangat penting untuk deteksi dini. Histopatologi pascanatal mengonfirmasi limfangioma dengan menunjukkan saluran limfatik yang melebar tanpa komponen solid. Kasus ini menekankan pentingnya manajemen multidisiplin dan pencitraan serial untuk memperjelas prognosis.
Kesimpulan: Diagnosis prenatal sangat penting dalam perencanaan manajemen. Ultrasonografi serial dan MRI dapat membantu memperjelas prognosis. Pendekatan multidisiplin sangat diperlukan untuk perawatan optimal.

Kata kunci: Diagnosis prenatal, Limfangioma, Malformasi limfatik, Manajemen perinatal, Massa ekstremitas janin.

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Introduction

Lymphangioma is a rare congenital malformation of the lymphatic system, resulting from abnormal transformation of lymph sacs during embryogenesis. The incidence of lymphangioma is approximately 1 in 6,000 pregnancies, with only 2% of cases occurring on the limbs. These lesions are characterized by dilated vascular spaces involving both the superficial and deeper dermal layers, often associated with malformed lymphatic vessels. While the exact etiology remains unclear, lymphangiomas are believed to arise due to the failure of primitive lymphatic channels to connect with the venous system, leading to lymph accumulation and cyst formation.[1,2]

Prenatal diagnosis of fetal lymphangioma is rare, particularly when presenting as a large mass on the limb, and it can be challenging to differentiate from other congenital anomalies such as sacrococcygeal teratoma or hemangioma. Advances in ultrasonography and fetal imaging have improved early detection, enabling better prenatal counseling and management planning.[3]

Case Presentation

A 39-year-old woman, G6P4A1L4, was referred to the Fetomaternal Department of Arifin Achmad Hospital at 25+6 weeks of gestation due to concerns about progressively increasing fetal limb size and fetal ascites detected during a routine ultrasound. The patient had no significant medical or family history of congenital abnormalities. Her prenatal screening tests were unremarkable. She first noticed an abnormal fetal limb shape during a routine antenatal visit at 21 weeks, but no prior ultrasound confirmed the presence of a mass at that time.

Ultrasound examination revealed a single live intrauterine fetus with an estimated fetal weight of 1,319 grams, in a head-down position. The anatomical assessment was normal, except for a 10 cm × 8 cm × 6 cm multicystic, hypoechoic tumor mass involving the entire left leg and left buttock. Initially, the mass was suspected to be a sacrococcygeal teratoma due to its rapid growth and extensive involvement. The presence of fetal ascites raised concerns

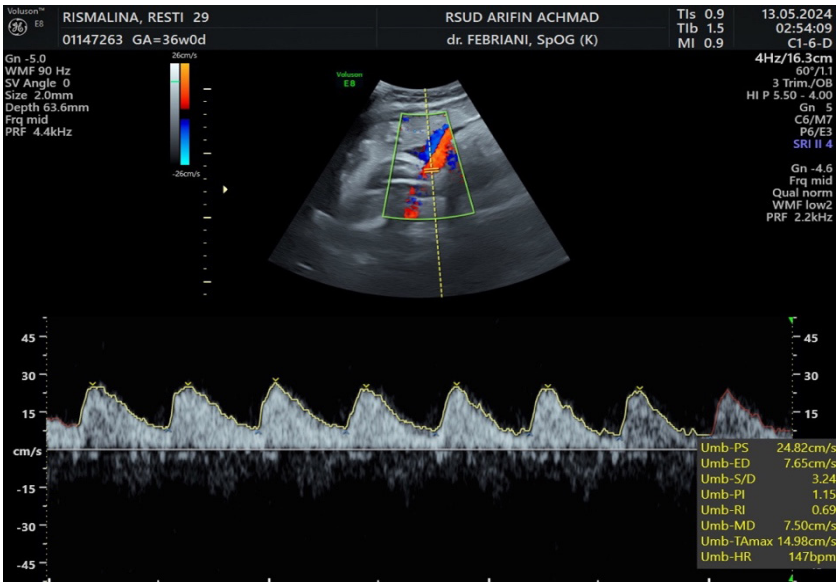

Figure 1 Ultrasound screening examination showed a hypoechoic mass in the left thigh of the fetus extending to the left buttock.

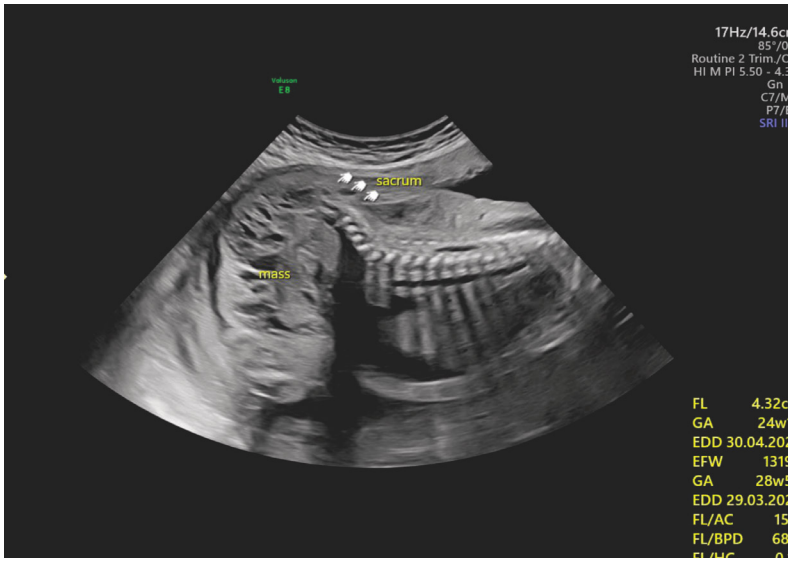

Figure 2 Umbilical artery Doppler examination showed within normal limits.

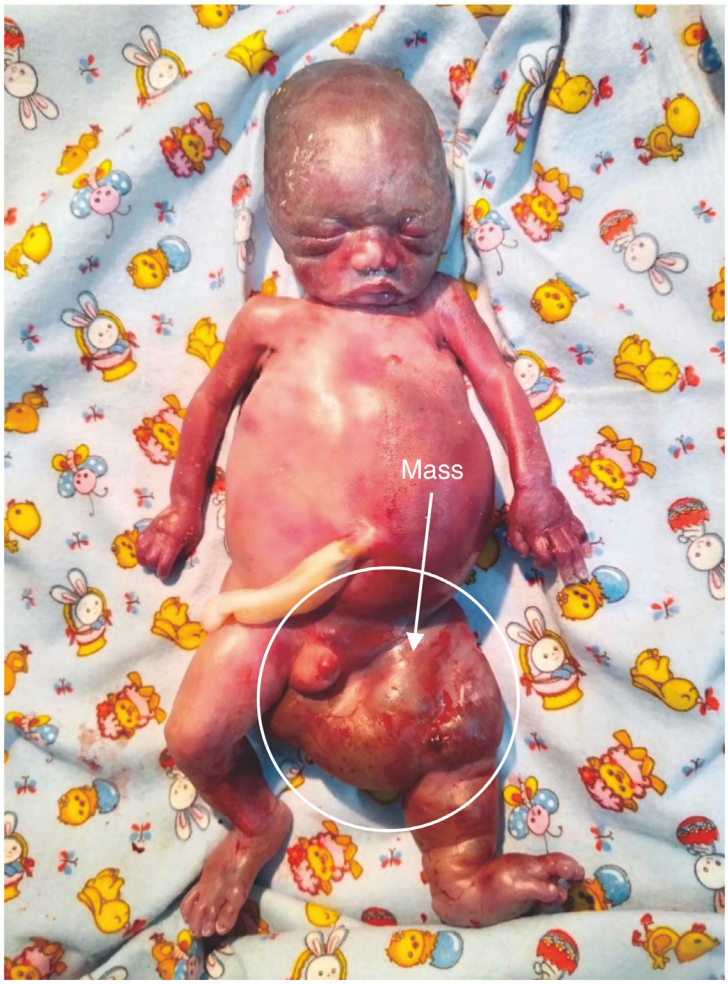

Figure 3. Clinical Finding A Large Mass Of Baby Leg.

about lymphatic obstruction and systemic fluid imbalance.

Doppler ultrasound findings showed reduced vascularity within the lesion, supporting a lymphatic origin rather than a highly vascularized tumor such as a teratoma or hemangioma. Given the poor prognosis associated with the tumor's progression, the parents elected to terminate the pregnancy after thorough counseling and an ethics committee review involving a multidisciplinary team. As there were no contraindications to vaginal delivery, labor was induced with 50 mcg of oral misoprostol every four hours, administered twice, leading to delivery. Four hours after the initiation of labor induction, a baby boy was delivered, weighing 1,400 grams, with an APGAR score of 3/0. The baby's left thigh was significantly enlarged, with prominent blood vessels visible. Despite resuscitation efforts, the baby was pronounced dead 15 minutes after birth. The mother's postpartum condition remained stable, with normal vital signs and laboratory results.

Investigation

Ultrasonography findings demonstrated a well-defined, multicystic mass affecting the left thigh, extending to the buttock region. The cystic structures varied in size and measured 10 cm × 8 cm × 6 cm. Doppler ultrasound findings revealed minimal vascularity, further supporting the lymphatic origin.

The presence of fetal ascites raised additional concerns about lymphatic obstruction and fluid imbalance. After delivery, a biopsy was performed to confirm the nature of the lesion. Histopathological examination revealed multiple dilated lymphatic channels lined by flattened endothelial cells, consistent with lymphangioma. The absence of solid components or immature tissues ruled out sacrococcygeal teratoma. Laboratory results, including maternal blood tests, were within normal limits.

Differential Diagnosis

Several congenital anomalies can mimic lymphangioma, making differential diagnosis essential:

1. **Sacrococcygeal Teratoma (SCT)**
   A germ cell tumor presenting as a cystic or solid mass in the sacrococcygeal region. Unlike lymphangioma, SCT often exhibits calcifications and solid components on imaging.[4,5]
2.. **Hemangioma**
   A benign vascular tumor that may appear as a soft-tissue mass with high vascularity on Doppler ultrasound.[3,4]
3. **Cystic Hygroma**
. A type of lymphangioma usually found in the cervical region rather than the extremities, often associated with chromosomal anomalies such as Turner syndrome.[3,4]

Treatment and Management

Given the extensive growth of the tumor and the presence of ascites, the prognosis for this fetus was poor. The parents opted for termination of pregnancy, which was carried out vaginally. A preterm male newborn weighing 1400 g was delivered spontaneously with an Apgar score of 3/0. The neonate exhibited a large mass on the left thigh.

Postnatal management of lymphangiomas typically involves observation, surgical resection, or sclerotherapy, depending on the size, location, and complications associated with the lesion. In this case, histopathological confirmation ruled out more aggressive conditions, but further management would have been required had the pregnancy continued.

Outcome and Follow-Up

The neonate did not survive due to complications related to prematurity and extensive lymphatic malformation. If diagnosed earlier in gestation, continuous monitoring with serial ultrasound and fetal MRI could have provided additional prognostic information and aided in perinatal decision-making. Postnatal treatment strategies include surgical excision or sclerotherapy using agents such as bleomycin and picibanil, depending on lesion characteristics and associated symptoms.

Discussion

Lymphangiomas are rare congenital malformations of the lymphatic system, posing significant diagnostic and management challenges, particularly when they manifest in the lower limbs. The etiology of lymphangiomas is believed to be linked to the failure of embryonic lymphatic channels to establish proper connections with the venous system, leading to lymph accumulation and cyst formation. This case highlights the complexity of prenatal diagnosis, given the initial suspicion of a sacrococcygeal teratoma due to the mass's size and location. However, detailed ultrasonographic findings, including the multicystic nature and lack of solid components or vascularity, ultimately supported the diagnosis of lymphangioma.[6,7]

Prenatal detection of lymphangioma is essential for parental counseling and management planning. In this case, fetal MRI could have provided additional clarity in distinguishing between differential diagnoses such as hemangiomas and teratomas.[8,9] The presence of fetal ascites further complicated the prognosis, raising concerns about lymphatic obstruction and fluid imbalance. Despite the detailed assessment, the rapid progression of the mass led to a poor fetal outcome, reinforcing the importance of early screening and serial imaging in cases presenting with limb abnormalities. Termination of pregnancy was pursued following ethical discussions involving a multidisciplinary team comprising obstetricians, neonatologists, pediatric surgeons, and the medical ethics committee. The labor was induced using misoprostol, following FIGO guidelines (50 mcg orally every 4 hours, administered twice), resulting in vaginal delivery. Postpartum, the mother's condition remained stable with normal vital signs and laboratory results.[10]

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Conclusion

Prenatal diagnosis of lymphangioma is crucial for appropriate management planning and parental counseling. In cases of extensive involvement, serial ultrasound and fetal MRI can help refine diagnosis and guide decision-making. While lymphangiomas may be treated postnatally with surgical or sclerotherapy interventions, severe cases

with associated complications may result in poor perinatal outcomes. A multidisciplinary approach remains essential in managing such complex conditions, ensuring optimal maternal and fetal care.

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